











Diseases .

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S. S. Srinivasan

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more frequent in malarial regions. It is rather more common in females, and between the 30th and 50th years. The youngest patient was a child of eight months. It has followed a blow. Patients have had a tendency to hæmorrhage, but, as a rule, the disease appears in fairly healthy persons without any recognizable cause.

*Symptoms.*—Anæmia is not a necessary accompaniment of all stages of the disease; the subjects may look very healthy and well. The onset is insidious, and, as a rule, the patient seeks advice for progressive enlargement of the abdomen and shortness of breath, or the pallor, palpitation, and other symptoms of anæmia. Bleeding at the nose is common. Gastro-intestinal symptoms may precede the onset. Occasionally the first symptoms are of a very serious nature. In one case a boy played lacrosse two days before the onset of the final hæmatemesis; and in another case a girl, who had, it was supposed, only a slight chlorosis, died of fatal hæmorrhage from the stomach before any suspicion had been aroused as to the true condition.

The increase in the size of the *spleen* is the most prominent feature in the majority of the cases. Pain and tenderness are common, though the progressive enlargement may be painless. A creaking fremitus may be felt on palpation. The enlarged organ extends downward to the right, and may be felt just at the costal edge, or when large it may extend as far over the navel. In many cases it occupies fully one-half of the abdomen, reaching to the pubes below and extending beyond the middle line. As a rule, the edge, in some the notch or notches, can be felt distinctly. Its size varies greatly from time to time. It may be perceptibly larger after meals. A hæmorrhage or free diarrhoea may reduce the size. The pressure of the enlarged organ may cause distress after eating; in one case it caused fatal obstruction of the bowels. On auscultation a murmur may sometimes be heard over the spleen, and Gerhardt described a pulsation in it.

The long bones are tender; leukæmic tumors are rare but there may be tender localized swellings, particularly on the ribs, which yield to pressure.

The *pulse* is usually rapid, soft, compressible, but often full in volume. The veins may be large and full, and pulsation in those of the hand and arm is common. Toward the close œdema may occur in the feet or general anasarca. *Hæmorrhage* is common. There may be most extensive purpura or hæmorrhagic exudate into pleura or peritoneum. Epistaxis is the frequent form. Hæmoptysis, hæmaturia and intestinal hæmorrhage are not infrequently present. Bleeding from the gums may be present. Hæmatemesis proved fatal in four of our cases, and in a third a large cerebral hæmorrhage rapidly killed.

Local *gangrene* may develop, with signs of intense infection and fever. There are very few pulmonary symptoms. The shortness of breath is due, as a rule, to the anæmia. Toward the end there may be œdema of the lungs or pneumœmia. Gastro-intestinal symptoms are rarely absent, and vomiting are early features in some cases, and diarrhoea may be troublesome, even fatal. There may be a dysenteric process in the colon. Jaundice rarely occurs. Ascites may be a prominent symptom, probably due to the splenic tumor. A leukæmic peritonitis may occur, due to involvement of the membranes.

The nervous system is not often involved, and throughout the viscera. The Headache, dizziness, and fainting spells are present with the green tinge of the skin.

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Due to pressure of a leukæmic tumor on the cord. There is a peculiar *neuritis*, due chiefly to the extravasation of blood, but there may be aggregations of leucocytes, forming small leukæmic growths. Optic neuritis is rare. Deafness is frequent; it may appear early and possibly is due to hæmorrhage. Features like Ménière's disease may come on suddenly, due to leukæmic infiltration or hæmorrhage into the semi-circular canal.

The urine presents no constant changes; uric acid is always in excess. *Priapism* is present in many cases, sometimes at the onset. It may persist for weeks. The cause is thrombosis of the veins in some cases.

*Fever* was present in two-thirds of our series. Periods of pyrexia may alternate with prolonged intervals of freedom. The temperature may range from 102° to 103° F.

*Blood*.—In all forms of the disease the diagnosis must be made by the examination of the blood, as it alone offers distinctive features.

The striking change is an increase in the *leucocytes*. The average in our series was 298,700 per c. mm., and the average ratio to the red cells was 1 to 40. Counts above 500,000 c. mm. are common, and they may rise above 1,000,000 per c. mm. The proportion of white to red cells may be 1 to 5, or may even reach 1 to 1. There are instances in which the number of leucocytes exceeded that of the red corpuscles. The increase is in all the forms. The polynuclear neutrophiles make up from 30 to 50 per cent.; both the small and the large lymphocytes are increased; the eosinophiles and the mast cells show both a percentage and absolute increase. The abnormal cells, the myelocytes, range from 30 to 50 per cent. Normoblasts and megaloblasts are common. At first the red cell count may be normal, but sooner or later anæmia comes on, and the count may fall to 2,000,000 per c. mm. The color index is usually low. The blood plates are increased. Charcot-Leyden crystals may separate from the clots and the hæmoglobin shows a remarkable tendency to crystallize.

*ALUKÆMIC INTERVALS*.—It has long been known that the white cells may fall to normal or even below. In a case in the Johns Hopkins Hospital, the leucocytes diminished from 500,000 per c. mm. on Jan. 26 to 6,000 on Feb. 16, and throughout the greater part of March were as low as 2,000 per c. mm. This followed the use of arsenic. With this the spleen may or may not reduce. The same may occur spontaneously, but has been frequently seen following benzol, radium and X-ray treatment. The question arises whether it is always possible in the aleukæmic intervals to diagnose the disease from the examination of the blood. In some cases the films are normal. These aleukæmic phases unfortunately are only transitory.

**II. LYMPHOID LEUKÆMIA**.—Less common, this occurs in acute and chronic forms.

**A. ACUTE LYMPHATIC LEUKÆMIA** is the most terrible of all blood diseases. It occurs in younger persons and more frequently in males. In onset and course the disease resembles an acute infection. Swelling of the tonsils, ulcerative angina, stomatitis, fever, hæmorrhages and a rapid anæmia are the dominant features. Dyspnœa, nausea, vomiting, and diarrhœa are not infrequently present. The disease resembles fulminant purpura, and cutaneous hæmorrhages are common. The patient feels ill. The glands of the neck are enlarged, but death may occur without marked aden-

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itis. The spleen is usually palpable, rarely very large. Hæmorrhages into the mucous membranes, and into the serous sacs are common. The course is rapid, and death may occur within a week of onset; more often in from three to six weeks. Remissions may occur and a case beginning acutely may linger for three or four months.

*Leukæmia cutis*, most common in this form, is characterized by nodular tumors in the skin, which may break down rapidly, hæmorrhages, pigmentation of the skin, and fever. The spleen and lymph-glands may be little, if at all, enlarged.

The blood picture in the acute form may give the only data for diagnosis. The anæmia is rapid with the usual changes in the blood cells. The leucocytes are increased but less as a rule than in the myeloid forms. Counts of 100,000 to 200,000 per c. mm. are frequent and the count may rise to above 1,000,000. The distinctive feature is the predominance of large lymphocytes, usually over 90 per cent. Atypical blood pictures may be met with—a mixed small and large lymphocytosis, macrolymphocytes and their variants.

The enlargement of the spleen and lymph-glands is less marked than in the myeloid form. Lymphoid swellings in the mouth, throat and intestines are common, and small tumors may be widely scattered on the serous membranes, skin, in the lungs, and even in the nervous system. The bone-marrow is deep red, but the changes depend much on the duration of the disease.

B. CHRONIC LYMPHATIC LEUKÆMIA is less common. Its existence has been denied, but cases of three to thirteen years' duration have been reported. A patient of W. H. Draper's of New York seen ten years after the onset had a sheaf of blood counts from every clinician of note in Europe and the United States. There was no anæmia, the leucocytes were 242,000 per c. mm., the superficial lymph-glands were enlarged and the spleen of moderate size.

It occurs in older persons, rarely, if ever, in children; the general health may be very good and the only inconvenience felt is from the bunches of enlarged glands. The spleen is rarely very large; the mesenteric and retroperitoneal groups may form big tumors. After lasting two or more years acute symptoms may come on—fever, hæmorrhages, stomatitis, tonsillitis. Pigmentation of the skin, itching with urticaria and lymphomas may be present, giving a skin picture very like that of Hodgkin's disease. The blood shows at first little or no anæmia. The leucocytes are usually above 100,000 per c. mm. and very high counts are common. The small lymphocytes predominate up to 90-95 per cent. The large forms are rare until the late stages when anæmia supervenes and the other elements show little or no change.

III. ATYPICAL LEUKÆMIAS.—(1) *Mixed leukæmias*; in nearly all cases of myeloid leukæmia a certain percentage of lymphocytes is present, which toward the end may be increased.

(2) *Cases with atypical blood changes*, such as a very high percentage of eosinophiles, or a very high proportion of plasma cells.

(3) *Chloroma* is an atypical lymphoid leukæmia in which the lymphatic tumors have a greenish color. It is more common in children. Exophthalmos is frequent owing to tumor formation in the orbit. The tumor growths occur chiefly in the skull, orbit, long bones, and throughout the viscera. The typical picture of this distribution may be present without the green tint of

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**Leukæmia.** The nature of the pigment is unknown. The blood picture is that of leukæmia.

(4) *Leukanæmia*.—This term was invented by Leube to describe a condition showing features both of leukæmia and severe anæmia. Glandular enlargement is usually present; the onset may be like the acute types of leukæmia, and the blood picture either of the lymphoid or of the myeloid type.

**Diagnosis.**—The recognition of the acute forms may be difficult, particularly those which begin with marked angina and cutaneous hemorrhages. It may not be until a blood examination is made or the glands enlarge that suspicion is aroused. The chronic forms are easily recognized. The enlarged spleen at once suggests a blood count, upon which alone the diagnosis rests. The diagnosis may be made by the ophthalmic surgeon. In the lymphatic form, too, the diagnosis rests with the blood examination. One has to recognize that there are certain cases of sepsis with marked lymphocytosis, in which the white blood corpuscles may reach 30,000 or 40,000 per c. mm. When the regional lymph-glands are involved this may raise a doubt. Infectious mononucleosis may give difficulty; the absence of anæmia is important. Under treatment with arsenic, radium or X-rays the increase of leucocytes may disappear, but the differential count may still be characteristic.

**Prognosis.**—Recovery is practically unknown. The acute cases die within three months; the chronic forms last from six months to four or five years, or longer. The chronic lymphatic form is the most protracted.

**Association with Other Diseases.**—Tuberculosis is not uncommon. Dock collected 27 cases, in none of which did the tuberculosis show any special influence. Intercurrent infections as influenza, erysipelas, or sepsis may have a remarkable effect upon the disease. In a case reported by Dock, after an attack of influenza the leucocytes fell from 367,000 to 7,500 per c. mm. A course of antistreptococcic serum may do the same.

**Treatment.**—Fresh air, good diet, and abstention from mental worry and care are the important general indications. The *indicatio morbi* can not be met. No treatment seems to influence the acute forms. There are certain remedies which have an influence upon the chronic forms. Arsenic should always be given. Fowler's solution can be begun in doses of three drops and increased to the limit of tolerance or sodium cacodylate given by injection. Benzol has been used but should be given with caution and discontinued if there is a drop in the red-cell count. If the number of leucocytes decreases steadily, the drug should be discontinued when they fall to 25,000. The dose is 2 i (4 c. c.) per day given in capsules with olive oil. The X-rays and radium, while not curative, add materially to the duration of life. They should not be used in the acute forms. The exposure should be over the spleen and long bones and care should be taken to watch for any signs of toxæmia. They usually cause a marked drop in the leucocytes. Either may be used with arsenic or benzol administration. Removal of the spleen has been done after radium treatment but the value of this is doubtful. Recurrence is to be expected after any treatment.

## HODGKIN'S DISEASE

### III. HODGKIN'S DISEASE

**Definition.**—A disease characterized by enlargement of the lymph-glands with anæmia and a fatal termination.

Anatomically there is an increase in the adenoid tissue of the glands, proliferation of the endothelial cells, formation of mononuclear and multinuclear giant cells, the presence of eosinophiles, and thickening of the fibrous reticulum.

**History.**—In 1832 Hodgkin recorded a series of cases of enlargement of the lymphatic glands and spleen. From the motley group that Hodgkin described, Wilks picked out the disease and called it *anæmia lymphatica*. Other names that have been given to it are *adénie* by Trousseau, *pseudo-leukæmia* by Cohnheim, *generalized lymphadenoma* and *malignant lymphoma*.

**Etiology.**—A widely spread disease, a majority of the cases occur in young adults, and more frequently in males than in females. Twins and sisters have been known to be attacked. The cause is unknown. Certain features suggest an acute infection: the rapid course of some cases, the association with local irritation in the mouth and tonsils, the frequency with which the disease starts in the cervical glands, the gradual extension from one gland group to another, and the recurring exacerbations of fever. Bunting and Yates described a diphtheroid organism with which they produced in the monkey a chronic lymphadenitis clinically resembling Hodgkin's disease. Possibly the disease is a spirillosis—in favor of which are the eosinophilia, the presence of eosinophilic cells in the glands, and the influence of arsenic. Sternberg suggested that the disease was a special form of tuberculosis; but the histological changes are characteristic, tubercle bacilli are not present in uncomplicated cases, the tuberculin test may be negative, and when present tuberculosis appears to be a terminal infection.

**Morbid Anatomy.**—The superficial lymph-glands are extensively involved, and from the cervical groups they form continuous chains uniting the mediastinal and axillary glands. The masses may pass beneath the pectoral muscles and even beneath the scapula. Of the internal glands, those of the thorax are most often affected, and the tracheal and bronchial groups may form large masses. The trachea and the aorta with its branches may be completely surrounded: the veins may be compressed, rarely the aorta itself. The masses perforate the sternum and invade the lung deeply. The retroperitoneal glands may form a continuous chain from the diaphragm to the inguinal canals. They may compress the ureters, the lumbar and sacral nerves, and the iliac veins. They may adhere to the broad ligament and the uterus and simulate fibroids. At an early stage the glands are soft and elastic; later they may become firm and hard. Fusion of contiguous glands does not often occur, and they tend to remain discrete, even after attaining a large size. The capsule may be infiltrated and adjacent tissues invaded. On section the gland presents a grayish white semi-translucent appearance, broken by intersecting strands of fibrous tissue: there is no caseation or necrosis unless a secondary infection has occurred.

The spleen is enlarged in 75 per cent. of the cases; in young children the enlargement may be great, but the organ rarely reaches the size of the spleen.

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**Secondary leukaemia.** In more than half of the cases lymphoid growths are present. The marrow of the long bones may be converted into a rich lymphoid tissue. The lymphatic structures of the tonsillar ring and of the intestines may show marked hyperplasia. The liver is often enlarged, and may present scattered nodular tumors, which may also occur in the kidneys.

**Histology.**—The studies of Andrewes and of Dorothy Reed show a very characteristic microscopic picture—proliferation of the endothelial and reticular cells, with the formation of lymphoid cells of uniform size and shape, and characteristic giant cells, the so-called lymphadenoma cells, containing four or more nuclei. Eosinophiles are always present, and proliferation of the stroma leads to fibrosis of the gland. The difference between the soft and hard forms depends largely upon the stage. When tuberculosis occurs as a secondary infection the two processes may be readily distinguished.

**Symptoms.**—A tonsillitis may precede the onset. Enlargement of the cervical glands is usually an initial feature; it is rare to find other superficial groups or the deeper glands attacked first. Months or even several years may elapse before the glands in the axillæ and groin become involved. During what may be called the first stage the patient's general condition is good; then anæmia comes on, not marked at first, but usually progressive. In the majority of cases the spleen is enlarged, but it never reaches the dimension of the leukæmic organ. There may be very little pain until the internal glands become involved. The lymph-nodes of the thorax and abdomen are frequently affected. With swelling of the *mediastinal glands* there are cough, dyspnoea, and often intense cyanosis, with all the signs of intrathoracic tumor. There may be moderate fever. Bronzing of the skin may occur, apart from the use of arsenic. Pruritus may be present and boils and ecchymatous blebs may occur. The blood shows a secondary anæmia, sometimes with a moderate leucocytosis. The leucocytes show no characteristic changes. There may be a moderate eosinophilia and, as the anæmia progresses, nucleated red cells appear, and toward the end there are instances of a great increase in the lymphocytes. As the disease progresses there is marked emaciation with athenia, and sometimes anasarca. This represents the common course, but there are many variations, among which the following are the most common:

(a) An ACUTE FORM has been described. In one case beginning, like so many cases of lymphatic leukaemia, with angina, the whole course was less than ten weeks. Ziegler mentions two cases of death within a month.

(b) LOCALIZED FORM.—The enlargement may be localized to certain groups, in the neck, groin, retroperitoneum, or thorax. Some of these cases present great difficulty in diagnosis, particularly if there are febrile paroxysms with slight involvement of the external groups. The disease may be confined to one region for a year or more before there is any extension. The localized mediastinal group often presents a remarkable picture—pressure signs, pain, orthopnoea—and, unless there are other groups involved, or enlargement of the spleen, it may be difficult to make the diagnosis during life.

(c) WITH RELAPSING PYREXIA.—To this remarkable form Pel and afterward Ebstein called attention. MacNalty made a careful study of this remarkable syndrome. Relapsing pyrexia may occur in cases with involvement of the internal glands alone, or, more frequently, with a general involvement of all the groups. Following on a period of low pyrexia, or of normal or

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subnormal temperature, there is a steady rise occupying two or three days to a maximum, which may reach  $105^{\circ}$ . For about three days the temperature remains at a high level, and then there is a gradual fall by lysis occurring in about three days, and the temperature then becomes sub-normal" (M. Nalty). An afebrile period of ten days or two weeks then occurs, to be followed by another bout of fever. This may be repeated for many months. In one case the pyrexia lasted for exactly fourteen days for many successive paroxysms. During the fever the glands swell and may become tender. This febrile form may occur with involvement of the internal glands alone. In one patient whose cervical glands had been thoroughly removed there were typical Pel-Ebstein paroxysms, and we could find no enlarged glands, internal or external.

(d) LATENT TYPE.—Ziegler called attention to the importance of this form, in which anæmia, fever, and constitutional symptoms may be present with enlargement of the internal glands. In one case the retroperitoneal glands alone were involved. Symmers reported an instance in which the glands and the hilus of the liver were attacked.

(e) SPLENOMEGALIC FORM.—Enlargement of the spleen is present in a large proportion of cases. Whether or not there is a type involving the spleen without the lymph-glands is still a question. Formerly many cases of simple enlargement of the spleen with or without anæmia were spoken of as *pseudo-leukæmia splenica*. It is not improbable that the disease may originate in the lymphoid tissue of the spleen, and cases have been reported by Ziegler, Symmers, Warrington, and others. It is very difficult to distinguish such cases clinically from the early stages of splenic anæmia.

(f) LYMPHOGRANULOMATOSIS.—The skin lesions may be in the rare form of a true lymphogranulomatosis or show a wide variety of changes. Among these are: pruritus, urticaria, œdema, petechiæ and marked pigmentation.

(g) LYMPHADENIA OSSIUM *has been described*—in which there have been multiple bone tumors of the bone marrow and periosteum with enlargement of the glands and spleen. How far these should be grouped with Hodgkin's disease seems very doubtful.

**Diagnosis.**—(a) TUBERCULOSIS.—In the case of enlargement of the glands on one side of the neck in a young person, it is often not easy to determine whether the disease is tuberculosis or Hodgkin's disease. Two points should be decided. First, one of the enlarged glands should be excised and carefully studied. The histological changes in Hodgkin's disease differ markedly from those in tuberculosis. Secondly, tuberculin should be used if the patient is afebrile. In early tuberculous adenitis the reaction is prompt and decisive. In the later stages, when many groups of glands are involved and cachexia well advanced, the tuberculin reaction may be present in Hodgkin's disease, but even then the histological changes are distinctive. Other points to be noted are the tendency in the tuberculous adenitis to coalescence of the glands, adhesion to the skin, with suppuration, etc., and the liability to tuberculosis of the lung or pleura. There is a form of generalized tuberculous adenitis which occurs particularly in negroes and simulates Hodgkin's disease with enlargement of the gland groups in the neck, arms and axilla, never perhaps so much as in Hodgkin's disease, but firm, elastic masses. There is irregular remittent fever, not with periods of apyrexia; the course

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protracted, and at autopsy only the lymph-glands may be found enlarged.

(b) **LEUKÆMIA.**—The blood examination gives the diagnosis and difficulty arises only in the cases of leukæmia in which the leucocytes decrease for a time become normal. Histologically there are striking differences between the structure of the glands in the two conditions.

(c) **LYMPHO-SARCOMA.**—Clinically the case may resemble Hodgkin's disease very closely, and in the literature the two diseases have been confounded. The glands, as a rule, form larger masses, the capsules are involved, and adjacent structures are attacked, but this may be the case in Hodgkin's disease. Pressure signs in the chest and abdomen are more common in sarcoma. But the most satisfactory mode of diagnosis is by examination of a gland.

**Course.**—There are acute cases in which the disease advances rapidly and death follows in a few months but, as a rule, it lasts for two or three years. Remarkable periods of quiescence may occur, in which the glands diminish in size, the fever disappears, and the general condition improves. Even a large group of glands may almost completely disappear, or a tumor mass in the neck may subside while the inguinal glands are enlarging. Usually cachexia with anæmia and cedema precedes death. A fatal event may occur early from great enlargement of the mediastinal glands.

**Treatment.**—When the glands are small and limited to one side of the neck, operation should be advised; even when both sides of the neck are involved, if there are no signs of mediastinal growth, operation is justifiable. The course of the disease may be delayed, even if cure does not follow.

Radium and the X-rays do good in selected cases. Certainly the glands have been reduced in size, but there is no proof of a complete cure. Other local treatment of the glands seems to do but little good.

Arsenic is the only drug which has a positive value and in some cases the effects on the glands are striking. It may be given in the form of Fowler's solution in increasing doses or as sodium cacodylate (gr. i-ii, 0.06-0.12 gm.) intramuscularly or intravenously daily or every second day. Ill effects from the larger doses are rare. Quinine and iron are useful as tonics. For the pressure pains morphia should be given.

## IV. PURPURA

Strictly speaking, purpura is a symptom, not a disease; but under this term are conveniently arranged a number of affections characterized by extravasations of blood into the skin. At present a satisfactory classification can not be made. Some group all forms under the designation *hæmorrhagic diathesis*, believing that intermediate forms link the mild purpura simplex and the most intense purpura hæmorrhagica. For a full discussion see Pratt's article in our *System of Medicine*.

The purpuric spots vary from 1 to 3 or 4 mm. in diameter. When small and pin-point-like they are called petechiæ; when large, they are known as ecchymoses. At first bright red in color, they become darker, and gradually fade to brownish stains. They do not disappear on pressure.

The following is a provisional grouping of the cases:

## PURPURA

**Symptomatic Purpura.**—(a) **INFECTIOUS.**—In pyæmia, septicæmia, malignant endocarditis (particularly in the last affection), ecchymata may be very abundant. In typhus fever the rash is always purpuric. In scarlet fever, and more particularly small-pox and cerebro-spinal fever, each a variety characterized by an extensive purpuric rash.

(b) **TOXIC.**—The venom of snakes produces extravasation of blood with great rapidity—a condition carefully studied by Weir Mitchell. Certain medicines, particularly copaiba, quinine, belladonna, mercury, ergot, and iodides may be followed by a petechial rash. Purpura may follow the use of small doses of iodide of potassium. A fatal event may be caused by a small amount, as in a case reported by Stephen Mackenzie of a child who died after a dose of  $2\frac{1}{2}$  grains. An erythema may precede the hæmorrhage. It is not always a simple purpura but may be an acute febrile eruption of great intensity. Workers with benzol, used as a solvent for rubber, may have severe purpura. Cases such as those reported by Selling have been in connection with the coating of tin cans, while the Swedish cases occurred in the manufacture of bicycle tires. Under this division comes the purpura associated with jaundice.

(c) **CACHECTIC.**—Under this heading are best described the instances of purpura which occur in cancer, tuberculosis, Hodgkin's disease, nephritis, scurvy, and in the debility of old age. In these cases the spots are usually confined to the extremities. They may be very abundant on the lower limbs and about the wrists and hands. This constitutes the commonest variety and many examples can be seen in the wards of any large hospital.

(d) **NEUROTIC.**—One variety is met with in cases of organic disease, the so-called myelopathic purpura, seen occasionally in tabes dorsalis, particularly following attacks of the lightning pains and, as a rule, involving the area of the skin in which the pains are most intense. Cases occur in acute myelitis and occasionally in severe neuralgia. Another form is the remarkable hysterical condition in which stigmata, or bleeding points, appear upon the skin.

(e) **MECHANICAL.**—This is most frequent with venous stasis, as in the paroxysms of whooping cough, in epilepsy and about tight bandages.

**Arthritic Purpura.**—This form is characterized by involvement of the joints. It is usually known, therefore, as "rheumatic," though in reality the evidence for this is not conclusive. Of 200 cases of purpura analyzed by Stephen Mackenzie, 61 had a history of rheumatism. It seems more satisfactory to use the designation arthritic. Three groups may be recognized.

(a) **PURPURA SIMPLEX.**—A mild form, often known as *purpura simplex*, is most common in children, in whom, with or without articular pain, purpuric spots appear upon the legs, less commonly upon the trunk and arms. As pointed out by Graves, this form may be associated with diarrhoea. The disease is seldom severe. There may be loss of appetite and slight anaemia. Fever is not, as a rule, present, and the patients get well in a week or ten days. Usually regarded as rheumatic, and associated, in some instances, with arthritic manifestations, yet in a majority the arthritis is slighter than in rheumatic fever and no other manifestations are present. The average duration is two to four weeks, but there are chronic cases lasting a year or more.

(b) **PURPURA (PELLOSIS) RHEUMATICA (Schönlein's Disease).**—This is characterized by multiple arthritis and an eruption which varies greatly in

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ter, sometimes *purpuric*, more commonly associated with *urticaria* or *erythema exudativum*. The purpuric spots are of small size and appear in successive crops. The disease is most common in males between the ages of twenty and thirty. It not infrequently sets in with sore throat, a fever from  $101^{\circ}$  to  $103^{\circ}$ , and articular pains. The rash, which appears first on the face or about the affected joints, may be a simple purpura or show ordinary articular wheals. In other instances there are nodular infiltrations, not to be distinguished from *erythema nodosum*. The combination of wheals and purpura, the *purpura urticans*, is very distinctive. Much more rarely vesication is met with, the so-called *pemphigoid purpura*. The amount of oedema is variable; occasionally it is excessive. These are the cases described as *febrile purpuric oedema*. The temperature range, in mild cases, is not high, but may reach  $102^{\circ}$  or  $103^{\circ}$  F.

The urine is sometimes reduced in amount and may be albuminous. The joint affections are usually slight, though associated with much pain, particularly as the rash comes out. Relapses may occur and the disease may return at the same time for several years in succession.

The diagnosis of this form rarely offers difficulty. The association of multiple arthritis with purpura and urticaria is very characteristic.

It is thought by most writers to be of rheumatic origin, and certainly some of the cases have the characters of rheumatic fever, *plus* purpura. By many, however, it is regarded as a special affection, of which the arthritis is a manifestation analogous to that which occurs in hamophilia and scurvy. The frequency with which sore throat precedes the attack, and the occasional occurrence of endocarditis or pericarditis, are suggestive of rheumatic fever.

The patients usually do well, and a fatal event is extremely rare. The throat symptoms may persist and give trouble. In some instances necrosis and sloughing of a portion of the uvula have followed.

**VISCERAL LESIONS IN PURPURA.**—In any form of purpura, in the erythemas, and in urticaria *visceral* lesions may occur. (a) *Gastro-intestinal crises*, pain, vomiting, melana, and diarrhoea. The attacks have often been mistaken for appendicitis or intussusception, and at operation the condition has been found to be an acute sero-hamorrhagic infiltration of a limited area of the stomach or bowel. Identical attacks occur in angio-neurotic oedema. Intussusception has occurred with purpura. These crises may occur for years in children before an outbreak of purpura or urticaria gives a clue to their nature. (b) Enlargement of the *spleen* is usually present in these cases. (c) *Albuminaria* and acute nephritis may occur and form the most serious complication, of which seven cases in the series died (*Am. J. Med. Sc.*, Jan., 1904). The combination of purpura with colic is usually spoken of as Henoch's purpura.

**Chronic Purpura.**—For years patients may have outbreaks of purpura without serious symptoms. One patient was practically never free from spots somewhere on the skin for thirty-three years, during which time she had had several severe attacks of nose-bleed, during which the purpura increased greatly. Another patient had recurring purpura on the legs for many years, with great pigmentation and thickening of the skin. There is a form of intermittent purpura with attacks over long series of years, as long as twenty,

## PURPURA

sometimes only on the skin, at other times with involvement of the mucous membranes (Elsner).

**Purpura hæmorrhagica.**—Under this heading (or thrombocytopenia) come cases of very severe purpura with hæmorrhages from the mucous membranes. The affection, *morbus maculosus* of Werlhof, is most common in young and delicate individuals, particularly in girls; but the disease may attack vigorous adults. After a few days of weakness and debility, purpuric spots appear on the skin and rapidly increase in number and size.

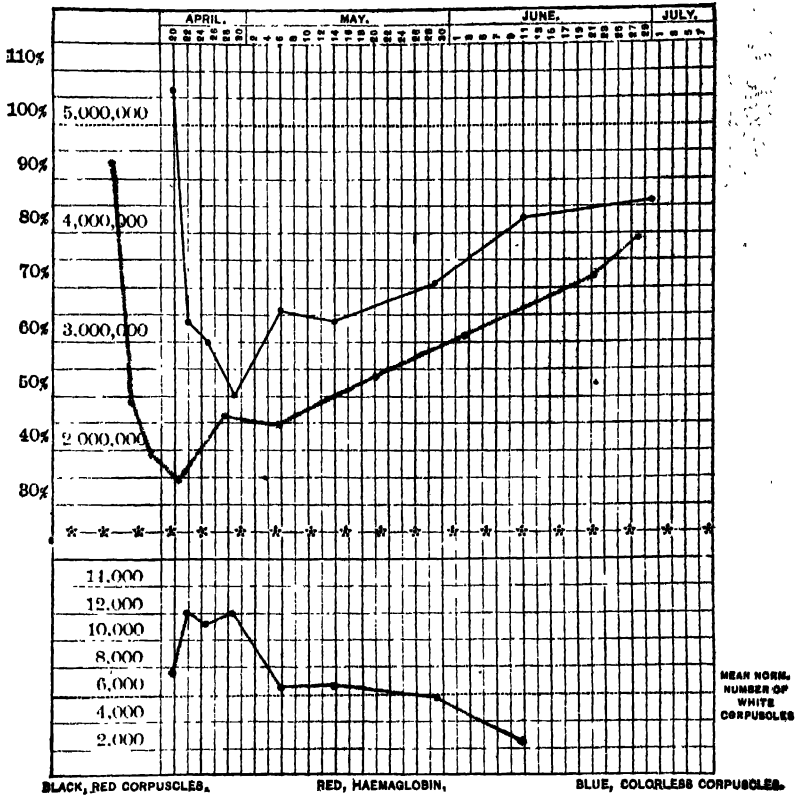


CHART XVI.—THE RAPIDITY WITH WHICH ANÆMIA IS PRODUCED IN PURPURA HÆMORRHAGICA, AND THE GRADUAL RECOVERY.

Bleeding from the mucous surfaces sets in, and epistaxis, intestinal hæmorrhage, hæmaturia, and hæmoptysis may cause profound anæmia. Death may take place from loss of blood or from hæmorrhage into the brain. Slight fever usually accompanies the disease. In favorable cases the course is from ten days to two weeks, but the average duration is two months and there are chronic forms which persist for years. There are instances of purpura hæmorrhagica of great malignancy, which may prove fatal within twenty-four hours—*purpura fulminans*. This form is most common in children, is characterized chiefly by cutaneous hæmorrhages, and death may occur before any bleeding takes place from the mucous membranes.

In the diagnosis of purpura hæmorrhagica it is important to exclude

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which may be done by the previous history, the circumstances under which the disease occurs, and by the absence of swelling of the gums. The malignant forms of the fevers, particularly small-pox and measles, are distinguished by the prodromes and the higher temperature. As regards the special blood features, the blood plates are markedly decreased, there is prolonged bleeding time and a non-retractile soft blood clot. In the other purpuras the blood plates are normal. The special points in the diagnosis from hæmophilia are considered under that disease. The possibility of mistaking the acute forms of *leukæmia* for purpura should be kept in mind.

**Treatment.**—In symptomatic purpura attention should be paid to the conditions under which it occurs, and measures should be employed to increase the strength and to restore a normal blood condition. Tonics, good food, and fresh air meet these indications. The patient should always be at rest in bed. In the simple purpura of children, or that associated with articular trouble, arsenic in full doses should be given. No good is obtained from the small doses, but Fowler's solution should be pushed freely until physiological effects are obtained. In *peliosis rheumatica* sodium salicylate may be given with discretion. It does not seem to have any special control over the hæmorrhages.

Aromatic sulphuric acid (Mxxv-xxx, 1-2 c. c.) may be given three times a day, but oil of turpentine is perhaps the best remedy, in 10 or 15-minim (1 c. c.) doses three or four times a day. The calcium salts, preferably the lactate, may be given in doses of 15 grains (1 gm.) three or four times a day for a few days. In bleeding from the mouth and nose the inhalation of carbon dioxide, irrigations with 2 per cent. gelatin solution, and epinephrine should be tried. The last remedy has often acted promptly. The treatment of the severe forms is the same as that given in hæmophilia. The intramuscular injection of 20-40 c. c. of citrated blood is a useful measure in severe cases. Splenectomy has been successful in purpura hæmorrhagica.

## HÆMORRHAGIC DISEASES OF THE NEW-BORN

**Syphilis Hæmorrhagica Neonatorum.**—The child may be born healthy, or there may be signs of hæmorrhage at birth. Then in a few days there are extensive cutaneous extravasations and bleeding from the mucous surfaces and from the navel. The child may become deeply jaundiced. The post mortem shows numerous extravasations in the internal organs and extensive syphilitic changes in the liver and other organs.

**Epidemic Hæmoglobinuria (Winkel's Disease).**—Hæmoglobinuria in the new-born, which occasionally occurs in epidemic form in lying-in institutions, is a very fatal affection, which sets in usually about the fourth day after birth. The child becomes jaundiced, and there are marked gastro-intestinal symptoms, with fever, jaundice, rapid respiration, and sometimes cyanosis. The urine contains albumin and blood coloring matter—methæmoglobin. The disease has to be distinguished from the simple icterus neonatorum, with which there may sometimes be blood or blood pigment in the urine. The post mortem shows an absence of any septic condition of the umbilical vessels, but the spleen is swollen and there are punctiform hæmorrhages in different parts. Some cases have shown marked acute fatty degeneration of the internal organs—the so-called Buhl's disease.

## HÆMOPHILIA

**Morbus maculosus Neonatorum.**—Apart from visceral hæmorrhages, result of injuries at birth, bleeding from one or more of the surfaces is uncommon in the new-born, particularly in hospital practice. Fifty occurred in 6,700 deliveries (C. W. Townsend). The bleeding may be from the navel alone, but more commonly it is general. Of Townsend's cases, 20 the blood came from the bowels, in 14 from the stomach, in 14 from the mouth, in 12 from the nose, in 18 from the navel, in 3 from the navel alone. The bleeding begins within the first week, but in rare instances is delayed to the second or third. Thirty-one of the cases died and 19 recovered. The disease is usually of brief duration, death occurring in from one to seven days. The temperature is often elevated. The nature of the disease is unknown. As a rule, nothing abnormal is found post mortem. The general nature of the affection, its self-limited character, the presence of fever, and the greater prevalence in hospitals suggest an infectious origin (Townsend). The bleeding may be associated with intense hæmatogenous jaundice. Not every case of bleeding from the stomach or bowels belongs in this category. Ulcers of the œsophagus, stomach, and duodenum have been found in the new-born. The child may draw blood from the breast and subsequently vomit it.

**Treatment.**—The most useful measure is the intramuscular injection of fresh or citrated human blood in amounts of 20-40 c. c. This should be repeated every four to eight hours if the hæmorrhage continues.

## V. HÆMOPHILIA

**Definition.**—A disease characterized by deficiency in the thromboplastic substances, thereby rendering the individual liable to severe and recurring hæmorrhages. The defect is hereditary, confined to the male sex but transmitted by the female alone.

**History.**—Our knowledge of this remarkable condition dates from 1803, when John C. Otto, a Philadelphia physician, published "an account of an hæmorrhagic disposition occurring in certain families," and first used the word "bleeder." The works of Grandidier and of Wickham Legg give full clinical details, and the monograph of Bulloch and Fildes (Dulan & Co., London, 1911) presents in extraordinary detail every aspect of the disease.

**Distribution.**—A majority of the cases have been reported from Germany, Switzerland, and the United States. Jews are supposed to be more prone to the disease, but this Bulloch doubts, and he discredits the negro cases.

**SEX.**—Bulloch and Fildes claim to have established the fact of immunity in females, denying the authenticity of all the published cases (19). "In none of the families of bleeders . . . do we find any unequivocal evidence of abnormality in the women, that is to say, any abnormality beyond what might be expected in any collection of females taken at random."

**INHERITANCE.**—Otto pointed out in his original paper that while the females do not themselves bleed they alone transmit the tendency. Of 171 recorded instances of transmission, 160 conform to the "law of Nasse" that the disease is transmitted by the unaffected females—"the conductor" (Bulloch and Fildes). They explain the 11 exceptions, and conclude that the disease is not capable of being propagated through a male. Hæmophilia with-

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Demonstrable inheritance is very rare. It is the best illustration in man of sex-limited inheritance, the mechanism of which has been worked out so beautifully by Morgan and his pupils in *Drosophila*.

**Pathogenesis.**—The blood looks normal. Delay in the coagulation time, up to 30 or even 40 minutes, and imperfect clot formation are the outstanding features. In contrast to purpura hæmorrhagica the platelets are normal. The essential defect is a congenital inability to produce a proper thrombin, through the agency of which the fibrinogen is converted into fibrin. Sahli first suggested that the disease was due to a deficiency in the thrombokinase. "It may be classed as one of the ferment-deficiency diseases, with a strong hereditary association similar to other ferment-deficiency diseases such as cystinuria, alkaptonuria, etc." (Vines). The deficiency is relative, not absolute, and is on the organic side of the clotting mechanism, and not in the inorganic side, e. g., due to lack of calcium salts. One of the difficulties in explaining the bleeding in hæmophilia is the fact that the hæmorrhage continues in spite of the presence of clots in and about the wound. Addis believes that a higher amount of thrombokinase is required to produce rapid clotting in hæmophilic than in normal blood. In a wound, coagulation may occur only in those parts, as at the side, where the concentration of this material is highest; but the clot itself prevents the addition of further quantities of the thrombokinase from the tissues, and when the quantity of thrombin set free from the primary clot is insufficient completely to coagulate the blood in the centre of the wound, the bleeding may continue indefinitely.

**Symptoms.**—"The cardinal symptoms are three in number . . . an inherited tendency in males to bleed" (Bulloch and Fildes). A trifling injury, of no moment in a normal person, determines a hæmorrhage, which has no tendency to stop, but the blood trickles or oozes until death follows or there is arrest. The bleeding may be external, internal, or into joints. A majority of the attacks may be traced to trauma, but spontaneous bleeding may occur. The liability is first noticed in early childhood and persists to adult life, gradually diminishing and eventually disappearing, sometimes about puberty. Tooth extraction is a common cause. Epistaxis is frequent, heading the list in Grandidier's series of 331 cases. Other localities were: mouth 43, stomach 15, bowels 36, urethra 16, lungs 17, and a few instances of bleeding from the tongue, finger tips, tear papilla, eyelids, external ear, vulva, navel, and scrotum. Trivial operations, as circumcision, have been followed by fatal hæmorrhage. Abdominal colic, due to bleeding into the intestinal wall, may occur as in Henoch's purpura. The diagnosis of appendicitis may be made.

**Hæmarthrosis**, due to bleeding from the synovial membrane, and periarticular bleedings are common. The onset is rapid with marked swelling and a variable amount of pain. The knee or elbow is most commonly attacked, and the affection has been mistaken for tuberculosis or for an acute septic infection. König distinguishes three stages—hæmarthrosis, panarthrititis, and deformity.

**Prevention.**—The women of bleeder families should not marry or marrying, they should not bear children. Males may marry safely.

**Diagnosis.**—The monograph by Bulloch and Fildes should be read by all who value accuracy of observation and investigation. Forms of bleeding are

## POLYCYTHÆMIA VERA, ERYTHRÆMIA

so common that it is a simple matter to construct a pedigree showing an inherited "hæmorrhagic diathesis." It is essential for the diagnosis that an individual should have been more or less subject to bleeding from various parts throughout his life. "No solitary hæmorrhage, however inexplicable, should, in our opinion, be regarded as hæmophilia; it is necessary to show that the individual has been repeatedly attacked, if not from birth, from infancy" (Bulloch and Fildes). There is no method by which we can determine the deficiency of the organic ferment on which the bleeding depends.

In the diagnosis from purpura hæmorrhagica the following points are important. In hæmophilia puncture of the skin rarely causes hæmorrhage, in purpura it usually does; the number of blood plates is normal in hæmophilia, much reduced in purpura; the coagulation time is prolonged in hæmophilia (but not constantly so; it may be normal between attacks), normal or nearly so in purpura; the "bleeding time" is not prolonged in hæmophilia, much prolonged in purpura; in hæmophilia the blood clot retracts normally but not in purpura; the application of a tourniquet to the upper arm is without result in hæmophilia but in purpura results in the formation of petechiæ on the forearm. As regards heredity, it is well to remember that there are cases of hereditary purpura, some being found in hæmophilic families.

In *prevention* every effort should be made to avoid trauma and active games are forbidden. Operations should be avoided if possible. The giving of calcium lactate and thymus gland extract at intervals has been advised.

**Treatment.**—Treatment consists in an attempt to supply the missing substance by the injection of serum or transfusion. A most useful measure is the subcutaneous or intramuscular injection of fresh or citrated human blood in doses of 20 to 40 c. c. Previous testing is not necessary. Fresh blood or serum from animals, such as the horse or rabbit, is also effective given subcutaneously in the same dosage. The injection should be repeated every twelve hours while necessary. The use of fresh anti-diphtheritic serum may be effectual. With obstinate bleeding and severe anæmia transfusion should be done. For surface hæmorrhage, compression should be employed combined with the application of various substances, as epinephrine (1 in 1000), cocaine (5 per cent.), or fresh blood or serum. The last has been injected into or around the wound with advantage. In arthritic hæmorrhage, operation is contraindicated but aspiration may be done for excessive effusion, as a needle puncture does not bleed as a rule. The joint should be kept at rest in the acute stage and passive motion begun as soon as this is over.

## VI. POLYCYTHÆMIA VERA, ERYTHRÆMIA

(*Vaquez' Disease, Osler's Disease*)

**Definition.**—A symptom-complex characterized by cyanosis, polycythæmia and splenic enlargement. It is possible that it is not a specific disease but a syndrome with a varied etiology and pathology. Lucas (1912) in a study of the subject pointed out the difficulty of distinguishing between primary and secondary polycythæmia. Some regard erythræmia as an affection of the erythroblastic tissues, analogous to the leukoblastic activity in leukaemia. In

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In cases the blood shows features both of erythræmia and myeloid leukæmia either condition may be primary. Warthin has drawn attention to "Perza's Disease or Syndrome" in which the features mentioned above were associated with syphilitic disease of the pulmonary arteries. In the cases with pulmonary arterio-sclerosis the resulting changes are regarded as compensatory.

**Pathology.**—We see polycythæmia as a *secondary* condition in high altitudes, in congenital heart disease, with marked loss of fluid and in emphysema of the lungs. The high altitude form is compensatory to lack of oxygen in the air and there is an increased activity of the bone marrow. In erythræmia proper an increased activity of the bone-marrow is present. The *blood* has a high uric acid content (probably endogenous); the iron content is high; the resistance of the corpuscles is increased; the viscosity is high; and the coagulation time is prolonged. The splenic enlargement is a secondary result of increased blood formation and destruction. In the cases with pulmonary arterio-sclerosis there is marked right heart hypertrophy.

**Symptoms.**—The three cardinal features are a change in the appearance of the patient, enlargement of the spleen, and polycythæmia. The superficial blood vessels, capillaries, and veins look full, so that the skin is always congested, in warm weather of a brick red color, in cold weather cyanosed. The engorgement of the face may be extreme, extending to the conjunctivæ, and in cold weather the cyanosis of the face and hands may be as marked as any that is ever seen. There is often marked vasomotor instability, the hand becoming deeply engorged when held down, and rapidly anæmic when held up.

The *spleen* is usually enlarged, but not to the great extent of leukæmia. It may vary in size from time to time. It is hard, firm, and painless.

The total bulk of *blood* is enormously increased, and the ratio of corpuscles to plasma is high. The polycythæmia ranges from 7 to 12 or even 13 millions of red cells per c. mm. As a rule, they are normal in appearance and shape; nucleated red cells may be present; the hæmoglobin ranges from 130 to 160 per cent., but the color index rarely reaches one. Moderate leucocytosis is the rule with a varying percentage of the different forms; a few myelocytes may be present. The specific gravity is high.

Of other symptoms the most common are incapacity for work, headache, flushing and giddiness. Constipation is common, and albuminuria is usually present. The blood pressure may be high; occasionally there are hæmorrhages into the skin and from the mucous membranes. Recurring ascites, probably in association with the splenic tumor, is present in some cases. There may be syncope and extreme prostration. Digestive disturbances may occur with pain of such severity that the abdomen has been explored. The cyanosis is intermittent in some cases.

Christian has emphasized the frequency of nervous symptoms, among which are headache, dizziness, paræsthesias, paresis and paralysis. Disturbances of vision are common. In some cases the symptoms suggest brain tumor. In early stages circulatory disturbance is probably responsible; later cerebral hæmorrhage or thrombosis occurs.

Morris reported three cases with the general appearance of the disease and with slight enlargement of the spleen, but without polycythæmia. Geisböck

## ENTEROGENOUS CYANOSIS

described a variety, *polycythæmia hypertonica*, with increased tension, arteriosclerosis, and nephritis.

In the form called "Ayerza's Disease" or "cardiacos negros," associated with syphilitic disease of the pulmonary arteries, there is headache, vertigo, somnolence, cyanosis, dyspnoea, cough, hæmoptysis, and polycythæmia. There is a pulmonary stage lasting for some years followed by the "cardiacos negros" stage lasting for two to five years, with marked enlargement of the right heart. The X-ray plate shows the shadow of the dilated pulmonary artery.

**Diagnosis.**—The features above referred to are sufficient in the absence of congenital heart disease, emphysema, and forms of cyanosis associated with poisoning by coal tar products. In a few rare cases the polycythæmia has been associated with tuberculosis of the spleen.

**Prognosis.**—The prognosis is bad for cure, but the condition may persist for years with reasonably good health. Cardiac failure, hæmorrhage, and recurring ascites have been the usual modes of death.

**Treatment.**—When there is much fullness of the head and vertigo, repeated bleedings have given relief. Inhalations of oxygen may be tried when the cyanosis is extreme. Saline purges and a diet low in purin and iron content are also helpful. The use of phenylhydrazin hydrochloride in doses of gr.  $1\frac{1}{2}$  (0.1 gm.) once daily, with a careful watch of the blood count, has proved useful. Benzol is of value in some cases. It can be given in doses of  $\mathfrak{m}$ . xv (1 c. c.) three times a day and the dose increased even to 3 i (4 c. c.). The blood count is a good guide for the proper dose. If syphilis is suspected, active treatment should be given. Radium and X-ray treatment over the long bones and spleen has apparently been useful in some cases, but we have not seen benefit from it. The number of leucocytes is a guide to the frequency of exposure; leucopenia should be avoided. Splenectomy should not be performed.

## VII. ENTEROGENOUS CYANOSIS

(*Methæmoglobinæmia and Sulphæmoglobinæmia*)

**Definition.**—A form of permanent cyanosis due to changes in the composition of the hæmoglobin of the blood.

**Etiology.**—It has long been known that certain drugs induced changes in the hæmoglobin. In poisoning by potassium chlorate methæmoglobinæmia occurs often with an active hæmolysis. Carbon monoxide, sulphuretted hydrogen, and some coal-tar products may cause a chronic cyanosis. Stokvis brought forward evidence to show that certain cases of chronic cyanosis are associated with intestinal disturbances, and he gives this form the name "enterogenous." Some are associated with methæmoglobinæmia, others with sulphæmoglobinæmia. Nitrite-producing organisms have been found which give a reducing substance which acts on hæmoglobin. In a doubtful case, with absence of lesions of the heart or lungs, a spectroscopic examination of the blood will determine if the cyanosis is of this nature, and which of the two derivatives of hæmoglobin is causing it.

In *methæmoglobinæmia* chronic diarrhœa is common sometimes associated with parasites. In Stokvis' case there was clubbing of the fingers without

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recognizable cause. Gibson and Douglas obtained from the blood of their patient a pure culture of a colon organism and suggested the name "microbic cyanosis." Methæmoglobinæmia has been met with in Winckel's disease, in one case of which a staphylococcus was isolated from the blood. A more striking combination is Boycott's discovery of an infective methæmoglo-minæmia in rats, caused by Gaertner's bacillus, which gives a remarkable bluish tint to the skin of white rats.

In *sulphæmoglobinæmia* the appearance of the patients is very much the same. They look very badly, even death-like, but feel comfortable, and there is no shortness of breath. The main complaints are cyanosis, constipation, weakness and headache. A nitrite-producing bacillus has been found in the saliva. Garrod suggests that it is a chronic poisoning by hydrogen sulphide, possibly absorbed from the intestines.

**Treatment.**—In some cases of methæmoglobinuria a milk diet has proved useful; meat and eggs should be excluded. Irrigation of the colon should be done systematically. In sulphæmoglobinæmia every effort should be made to keep the bowels moving freely. The use of a vaccine made from the nitrite-producing organism has proved successful. Foci of infection should be treated, especially in the mouth.

## SECTION X

# DISEASES OF THE CIRCULATORY SYSTEM

## A. DISEASES OF THE PERICARDIUM

### I. PERICARDITIS

Pericarditis is the result of infective processes, primary or secondary, or arises by extension of inflammation from contiguous organs. The *pus cocci*, the *pneumococcus*, and the *tubercle bacillus* are the chief causal organisms.

**Etiology.**—PRIMARY, so-called idiopathic, inflammation is rare; but it has occurred in children without any evidence of rheumatic fever or of local or general disease. Certain of the cases are tuberculous.

Pericarditis from *injury* usually comes under the care of the surgeon in connection with the primary wound. The trauma may be from within, due to the passage of a foreign body—a needle, a pin, or a bone—through the *oesophagus*—a variety exceedingly common in cows and horses.

**SECONDARY.**—(a) This occurs most frequently with *rheumatic fever*. In our 330 cases of rheumatic fever (Johns Hopkins Hospital) pericarditis occurred in twenty—practically 6 per cent. The articular trouble may be slight or the disease may be associated with acute tonsillitis in rheumatic subjects. Certain of the so-called idiopathic cases have their origin in an acute tonsillitis. The pericarditis may precede the arthritis. (b) In septic processes; in the acute necrosis of bone and in puerperal fever it is not uncommon. (c) In *tuberculosis*, in which the disease may be primary or part of a general involvement of the serous sacs or associated with extensive pulmonary disease. (d) In the *fevers*. Not infrequent after scarlet fever, it is rare in measles, small-pox, typhoid fever, and diphtheria. In pneumonia it is not uncommon, occurring in 31 among 665 cases (Chatard). In 184 post mortems there were 29 instances of pericarditis. It is most frequent in double pneumonia, and in our series with disease of the right side, if only one lung was involved. Pericarditis sometimes complicates chorea; it was present in 19 of 73 autopsies; in only 8 of these was arthritis present. (e) *Terminal pericarditis*. In gout, chronic nephritis, arterio-sclerosis, scurvy, diabetes, and chronic illness of all sorts a latent pericarditis is common and usually overlooked.

(f) *By Extension.*—In pneumonia it is most often met with in children and alcoholics. With simple pleurisy it is rare. In ulcerative endocarditis, purulent myocarditis, and in aneurism of the aorta pericarditis is occasionally found. It may also follow extension of infection from the mediastinal glands, the ribs, sternum, vertebræ, and even from the abdominal viscera.

Pericarditis occurs at all ages. Cases have been reported in the fetus. In the new-born it may result from septic infection through the navel. Throughout childhood the incidence of rheumatic fever and scarlet fever makes it a

## DISEASES OF THE CIRCULATORY SYSTEM

an affection, whereas late in life it is most often associated with tuberculosis, nephritis, and gout. Males are somewhat more frequently attacked than females. The so-called epidemics of pericarditis have been outbreaks of pneumonia with this as a frequent complication.

### ACUTE FIBRINOUS PERICARDITIS

This, the most common and benign form, is distinguished by the small amount of exudate which coats the surface in a thin layer and may be partial or general. In the mildest grades the membrane looks lustreless and roughened, due to a thin fibrinous sheeting, which can be lifted, showing beneath an injected or ecchymotic serosa. As the fibrinous sheeting increases in thickness the constant movement of the adjacent surfaces gives to it sometimes a ridge-like, at others a honeycombed appearance. With more abundant fibrinous exudation the membranes present an appearance like buttered surfaces which have been drawn apart. The fibrin is in shreds and the heart presents a curiously shaggy appearance—the hairy heart of old writers, *cor villosum*.

In mild grades the subadjacent muscle looks normal, but in the more prolonged and severe cases there is myocarditis, and for 2 or 3 mm. beneath the visceral layer the muscle presents a pale, turbid appearance. Some of these acute cases are tuberculous and the granulations are easily overlooked.

There is usually a slight amount of fluid entangled in the meshes of fibrin, but there may be very thick exudate without much serous effusion.

**Symptoms.**—Unless sought for there may be no objective signs, and for this reason it is often overlooked, and in hospitals the disease is relatively more common in the post-mortem room than in the wards.

**Pain** is a variable symptom, not usually intense, and in this form rarely excited by pressure. It is more marked in the early stage, and may be referred to the præcordia, to the region of the xiphoid cartilage or to the abdomen. The last may be so marked that the abdomen has been opened. In some instances the pain is of an aggravated and distressing character resembling angina. Cough and dyspnoea may occur. *Fever* is usually present, but it is not always easy to say how much depends upon the primary disease, and how much upon the pericarditis. It is as a rule not high, rarely exceeding 103.5° F. In rheumatic cases hyperpyrexia has been observed.

**PHYSICAL SIGNS.**—*Inspection* is negative; *palpation* may reveal a distinct fremitus caused by rubbing of the roughened pericardial surfaces. This is usually best marked over the right ventricle. It is not always to be felt, even when the friction sound on auscultation is loud and clear.

**Auscultation.**—The *friction* sound is one of the most distinctive of physical signs. It is double, corresponding to the systole and diastole; but the synchronism with the heart sounds is not accurate, and the to and fro murmur usually outlasts the time occupied by the first and second sounds. In rare instances the friction is single; more frequently it appears to be triple in character—a sort of canter rhythm. The sounds have a peculiar rubbing, grating quality, characteristic when once recognized, and rarely simulated by

## PERICARDITIS

endocardial murmurs. Sometimes instead of grating there is a *creaking* quality—the *bruit de cuir neuf*—the new leather murmur of the French. The pericardial friction appears superficial, very close to the ear, and is usually intensified by pressure with the stethoscope. It is best heard over the right ventricle, the part of the heart most closely in contact with the front of the chest—that is, in the fourth and fifth interspaces and adjacent portions of the sternum. There are instances in which the friction is most marked at the base, over the aorta, and at the superior reflection of the pericardium. Occasionally it is best heard at the apex. It may be limited to a narrow area, or transmitted up and down the sternum. There are no definite lines of transmission as in endocardial murmurs. An important point is the variability of the sounds, both in position and quality; they may be heard at one visit and not at another. The maximum of intensity will be found to vary with position. Friction may be present with a thin, almost imperceptible, layer of exudate; on the other hand it may not be present with a thick, buttery layer. The rub may be entirely obscured by the loud râles in pneumonia, in which disease pericarditis is recognized clinically in about half the cases.

**Diagnosis.**—There is rarely any difficulty in determining the presence of a dry pericarditis, for the friction sounds are distinctive. The double murmur of aortic insufficiency may simulate closely the to and fro pericardial rub. The constant character of the aortic murmur, the direction of transmission, the phenomena in the arteries, the blood pressure record, and the associated conditions should prevent this error.

*Pleuro-pericardial* friction is very common, and may be associated with endo-pericarditis, particularly in cases of pneumonia. It is frequent, too, in tuberculosis. It is best heard over the left border of the heart, and is much affected by the respiratory movement. Holding the breath or taking a deep inspiration may abolish it. The rhythm is not the simple to and fro diastolic and systolic, but the respiratory rhythm is superadded, usually intensifying the murmur during expiration and lessening it on inspiration. In tuberculosis of the lungs there are instances in which, with the friction, a loud systolic click is heard, due to the compression of a thin layer of lung and the expulsion of air from a softening focus or from a bronchus.

And, lastly, it is not very uncommon, in the region of the apex beat, to hear a series of fine crepitant sounds, systolic in time, often very distinct, suggestive of pericardial adhesions, but heard too frequently for this cause.

**Course.**—Simple fibrinous pericarditis never kills, but it occurs so often with serious affections that we have frequent opportunities to see all stages of its progress. In the majority of cases the inflammation subsides and the thin fibrinous laminae gradually become converted into connective tissue, which unites the pericardial surfaces firmly together. A very thin layer may “clear” without leaving adhesions. In other instances the inflammation progresses, with increase of the exudation, and the condition is changed from a “dry” to a “moist” pericarditis, or the pericarditis with effusion. In some instances the simple plastic pericarditis becomes chronic, and great thickening of both visceral and parietal layers gradually results.

## DISEASES OF THE CIRCULATORY SYSTEM

### PERICARDITIS WITH EFFUSION

**Etiology.**—Commonly a direct sequence of the dry or plastic pericarditis, of which it is sometimes called the second stage, this form is found most frequently in association with rheumatic fever, tuberculosis, and septicæmia, and sets in usually with præcordial pain, with slight fever or a distinct chill. In children the disease may, like pleurisy, come on without local symptoms, and, after a week or two of failing health, slight fever, shortness of breath, and increasing pallor, the physician may find, to his astonishment, signs of extensive pericardial effusion. These latent cases are often tuberculous. W. Ewart called special attention to latent and ephemeral pericardial effusions, which he thinks are often of short duration and of moderate size, with an absence of the painful features of pericarditis.

**Morbid Anatomy.**—The effusion may be sero-fibrinous, hæmorrhagic, or purulent. The amount varies from 200 to 300 c. c. to 2 litres. In the cases of sero-fibrinous exudation the pericardial membranes are covered with thick, creamy fibrin, which may be in ridges or honeycombed, or may present long, villous extensions. The parietal layer may be several millimetres in thickness and form a firm, leathery membrane. The hæmorrhagic exudation is usually associated with tuberculous or cancerous pericarditis, or with the disease in the aged. The lymph is less abundant, but both surfaces are injected and often show numerous hæmorrhages. Thick, curdy masses of lymph are usually found in the dependent part of the sac. In many cases the effusion is really sero-purulent, a thin, turbid exudation containing flocculi of fibrin.

The pericardial layers are greatly thickened and covered with fibrin. When the fluid is pus, they present a grayish, rough, granular surface. Sometimes there are distinct erosions on the visceral membrane. The heart muscle becomes involved to a greater or less extent and, on section, the tissue, for a depth of from 2 to 3 mm., is pale and turbid, and shows evidence of fatty and granular change. Endocarditis coexists frequently, but rarely results from the extension of the inflammation through the wall of the heart.

**Symptoms.**—Even with copious effusion the onset and course may be so insidious that no suspicion of the true nature of the disease is aroused.

As in the simple pericarditis, *pain* may be present, either sharp and stabbing or as a sense of distress and discomfort in the cardiac region. It is more frequent with effusion than in the plastic form. Pressure at the lower end of the sternum usually aggravates it. *Dyspnoea* is a common and important symptom, one which, perhaps, more than any other, excites suspicion and leads to a careful examination. The patient is restless, lies upon the left side or, as the effusion increases, sits up in bed. Associated with the dyspnoea is in many cases a peculiarly dusky, anxious countenance. The pulse is rapid, small, sometimes irregular, and may present the characters known as *pulsus paradoxus*, in which during each inspiration the pulse beat becomes very weak or is lost. These are due, in great part, to the direct mechanical effect of the fluid within the pericardium which embarrasses the heart's action. Other pressure effects are distention of the veins of the neck, dysphagia, which may be a marked symptom, and irritative cough from compression of the trachea. Aphonia is not uncommon, owing to compression or irritation of the recurrent laryngeal nerve. In massive effusion the pericardial sac occupies a large

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portion of the antero-lateral region of the left side and the condition is frequently mistaken for pleurisy. Even in moderate grades the left lung is somewhat compressed, an additional element in the production of the dyspnea. Great restlessness, insomnia, and in the later stages low delirium and coma are symptoms in the more severe cases. Delirium and marked cerebral symptoms are associated with the hyperpyrexia of rheumatic cases, but in addition there may be peculiar mental symptoms. The patient may become melancholic and show suicidal tendencies or the condition resembles closely delirium tremens. Sibson, who specially described the condition, states that the majority of such cases recover. Chorea may also occur, as was pointed out by Bright.

Convulsions are rare but have occurred during paracentesis.

**PHYSICAL SIGNS.**—*Inspection.*—In children the præcordia bulges and with copious exudation the antero-lateral region of the left chest becomes enlarged. A wavy impulse may be seen in the third and fourth interspaces, or there may be no impulse visible. The intercostal spaces bulge somewhat and there may be marked edema of the wall. Perforation externally through a space is very rare. Owing to compression of the lung, the expansion of the left side is greatly diminished. The diaphragm and left lobe of the liver may be pushed down and produce a distinct prominence in the epigastric region.

*Palpation.*—A gradual diminution and final obliteration of the cardiac impulse is a striking feature in progressive effusion. The position of the apex beat is not constant. In large effusions it is usually not felt. In children as the fluid collects the pulsation may be best seen in the fourth space, but this may not be the apex itself. The pericardial friction may lessen with the effusion, though it often persists with considerable fluid or it may be felt in the erect and not in the recumbent posture.

*Percussion* gives most important indications. The gradual distention of the pericardial sac pushes aside the margins of the lungs so that a large area comes in contact with the chest wall and gives a greatly increased area of dullness. The form of this dullness is irregularly pear shaped; the base or broad surface directed downward and the stem or apex directed upward. Shifting dullness at the base, that is a change in the width of the dull area with the patient recumbent and sitting, wider when recumbent, is an important sign. There is a disproportionate extension of dullness upward, and to the right, with dullness in the right fifth interspace, extending one or two inches to the right of the sternum (Roth's sign). Williamson could not verify this in an experimental study. In large effusions there may be altered resonance in the left axilla, and an area of dullness near the angle of the scapula with bronchial breathing, which may alter when the patient leans forward.

*Auscultation.*—The friction sound heard in the early stages may disappear when the effusion is copious, but often persists at the base or at the limited area of the apex. It may be audible in the erect and not in the recumbent posture. With the absorption of the fluid the friction returns. One of the most important signs is the gradual weakening of the heart sounds, which with the increase in the effusion may become so muffled and indistinct as to be scarcely audible. The heart's action is usually increased and the rhythm dis-

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**Inspected.** Occasionally a systolic endocardial murmur is heard. Early and persistent accentuation of the pulmonary second sound may be present.

**Important accessory signs** in large effusion are due to pressure on the left lung. The antero-lateral margin of the lower lobe is pushed aside and in some instances compressed, so that percussion in the axillary region, in and just below the transverse nipple line, gives a modified note, usually a dull tympany. Variations in the position of the patient may change the note in this area, over which on auscultation there is either feeble or tubular breathing. The left lobe of the liver is pushed down.

**Course.**—Cases vary extremely in the rapidity with which the effusion takes place. In every instance, when a pericardial friction murmur has been detected, the practitioner should outline with care the upper and lateral limits of cardiac dulness, mark the position of the apex beat, and note the intensity of the heart sounds. In many instances the exudation is slight in amount, reaches a maximum within forty-eight hours, and then gradually subsides. In other instances the accumulation is more gradual and progressive, increasing for several weeks. To such cases the term *chronic* has been applied. The rapidity with which a sero-fibrinous effusion may be absorbed is surprising. The possibility of the absorption of a purulent exudate is shown by the cases in which the pericardium contains semi-solid grayish masses in all stages of calcification. With sero-fibrinous effusion, if moderate in amount, recovery is the rule, with inevitable union of the pericardial layers. In some septic cases there is a rapid formation of pus and a fatal result may follow in three or four days. More commonly, when death occurs with large effusion, it is not until the second or third week and takes place by gradual asthenia.

**Prognosis.**—In the sero-fibrinous effusions the outlook is good, and a large majority of the rheumatic cases recover but with a damaged myocardium. The purulent effusions are more dangerous; the septic cases are usually fatal, and recovery is rare in the slow, insidious tuberculous forms. In nephritis, gout, acute infectious diseases and in old age the outlook is always grave.

**Diagnosis.**—Probably no serious disease is so frequently overlooked. Post-mortem experience shows how often pericarditis is not recognized, or goes on to resolution and adhesion without attracting notice. In a case of rheumatic fever, watched from the outset, with the attention directed daily to the heart, it is one of the simplest of diseases to diagnose; but when one is called to a case for the first time and finds perhaps an increased area of præcordial dulness, it is often very hard to determine with certainty whether or not effusion is present. The difficulty usually lies in distinguishing between dilatation of the heart and pericardial effusion. Although the differential signs are simple on paper, it is notoriously difficult in certain cases, particularly in stout persons, to say which condition exists. The special points are:

(a) The character of the impulse, which in dilatation, particularly in thin-chested people, is commonly visible and wavy. (b) The shock of the cardiac sounds is more distinctly palpable in dilatation. (c) The area of dulness in dilatation rarely has a triangular form; nor does it, except in cases of mitral stenosis, reach so high along the left sternal margin or so low in the fifth and sixth interspaces *without visible or palpable impulse*. An upper limit of dulness shifting with change of position speaks strongly for effusion. (d) In dilatation the heart sounds are clearer, often sharp or fetal in character; gal-

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lop rhythm is common, whereas in effusion the sounds are distant and muffled. (e) Rarely in dilatation is the distention sufficient to compress the lungs and produce a tympanitic note in the axillary region, or flatness behind. (f) The X-ray picture may be very definite.

The number of excellent observers who have failed to discriminate between these two conditions, and who have performed paracentesis *cordis* instead of paracentesis *pericardii*, is perhaps the best comment on the difficulties.

Massive (1½ to 2-litre) exudations have been confounded with a pleural effusion and the pericardium has been tapped under the impression that the exudate was pleuritic. The dull tympany in the infrascapular region, the absence of well-defined movable dulness, and the feeble, muffled sounds are indicative points. Followed from day to day there is rarely much difficulty, but it is different when a patient seen for the first time presents a large area of dulness in the left chest, and there is no pericardial friction murmur. Some cases have been regarded as encapsulated pleural effusions.

A special difficulty exists in recognizing the large exudate in pneumonia. The effusion may be much larger than the signs indicate, and the involvement of the adjacent lung and pleura is confusing. In at least three cases in our series we should have tapped the sac; post mortem the effusion was more than a litre.

The nature of the fluid can not be determined positively without aspiration; but a fairly accurate opinion can be formed from the primary disease and the general condition. In rheumatic cases the exudation is usually sero-fibrinous; in septic and tuberculous cases it is often purulent from the outset; in senile, nephritic, and tuberculous cases it may be hæmorrhagic.

**Treatment.**—The patient should have absolute quiet, mentally and bodily, so as to reduce the heart's action to a minimum. Drugs given for this purpose, such as aconite or digitalis, are of doubtful utility. The diet should be simple and liquids are usually best. The ice bag is of great value. It may be applied to the præcordia at first for an hour at a time, and then continuously. It reduces the frequency of the heart's action and seems to retard the progress of an effusion. Blisters are not indicated in the early stage but simple counter-irritation may give relief. Morphia should be given for pain or severe distress.

When *effusion* is present, the following measures to promote absorption may be adopted: Blisters to the præcordia, a practice not so much in vogue now as formerly. It is surprising, however, in some instances, how quickly an effusion will subside on their application. Purges and iodide of potassium are of doubtful utility. The action of the kidneys may be promoted by alkaline diuretics.

When signs of serious impairment of the heart occur, as indicated by dyspnoea, small, rapid pulse, dusky, anxious countenance, paracentesis or incision of the pericardium should be performed. With a sero-fibrinous exudate, aspiration is sufficient; but when the exudate is purulent, the pericardium should be freely incised and drained. The puncture may be made in the fifth or sixth interspace, outside the left nipple line. In large effusions the pericardium can be readily reached without danger by thrusting the needle upward and backward close to the costal margin in the left costo-xiphoid angle. With an earlier operation in many instances and a more radical one in others

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incision and free drainage, not aspiration, when the fluid is purulent—the percentage of recoveries will be greatly increased. Repeated tapping may be needed. One patient with tuberculous effusion, tapped three times, recovered and was alive three years afterward. In convalescence, the patient should be quiet for some time; the condition of the myocardium is a good indication as to the duration of rest.

### CHRONIC ADHESIVE PERICARDITIS

#### *(Adherent Pericardium, Indurative Mediastino-pericarditis)*

The remote prognosis in pericarditis is very variable. Some patients get well and have no further trouble, but in young persons serious results sometimes follow adhesions and thickening of the layers. As Sequira pointed out, the danger is directly in proportion to the amount of dilatation and weakening of the pericardium in consequence of the inflammation. The loss of the support afforded to the heart by the fibrous bag in which it is inclosed is an important factor. There are two forms of adherent pericardium.

(a) Simple adhesion of the peri- and epicardial layers, a common sequence of pericarditis, met with post mortem as an accidental finding. It is not necessarily associated with disturbance in the function of the heart, which in a large proportion of the cases is neither dilated nor hypertrophied.

(b) Adherent pericardium with chronic mediastinitis and union of the outer layer of the pericardium to the pleura and chest wall. This constitutes a most serious form of cardiac disease, particularly in early life, and may lead to an extreme grade of hypertrophy and dilatation. The peritoneum may be involved with perihepatitis, cirrhosis, and ascites (Pick's disease).

**Symptoms.**—The symptoms of adherent pericardium are those of hypertrophy and dilatation of the heart, and later of cardiac insufficiency (G. D. Head in a careful study of 59 cases divides them into (1) a small silent group with no symptoms, (2) a larger group with all the features of cardiac disease, and (3) a group comprising 11 cases in his series in which the features were hepatic. To this last group much attention has been paid since Pick's description. The *hepatic* features dominate the picture and the diagnosis of cirrhosis of the liver is usually made. Recurring ascites is the special feature and one patient was tapped 121 times. There is chronic peritonitis, with great thickening of the capsule of the liver and consequent contraction of the organ.

**Diagnosis.**—The following are important points in the diagnosis. *Inspection.* A majority of the signs of value come under this heading. (a) The *præcordia* is prominent and there may be marked asymmetry, owing to the enormous enlargement of the heart. (b) The extent of the cardiac impulse is greatly increased, and may sometimes be seen from the third to the sixth interspaces, and in extreme cases from the right parasternal line to outside the left nipple. (c) The character of the cardiac impulse. It is undulatory, wavy, and in the apex region there is marked systolic retraction. (d) Diaphragm phenomena. John Broadbent called attention to a very valuable sign. When the heart is adherent over a large area of the diaphragm there is with each pulsation a systolic tug, which may be communicated through the diaphragm to the points of its attachment on the wall, causing a visible

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retraction. This had long been recognized in the region of the seventh and eighth rib in the left parasternal line, but Broadbent called attention to the fact that it was frequently best seen on the left side behind, between the eleventh and twelfth ribs. This is a valuable and quite common sign, and may be very localized. One difficulty is that, as A. W. Tallant pointed out, it may occur in thin chested persons with great hypertrophy of the heart. Sir William Broadbent called attention to the fact that owing to the attachment of the heart to the central tendon of the diaphragm this part does not descend with inspiration, during which act there is not the visible movement in the epigastrium. The respiratory movement of the left thorax is decreased. (e) Diastolic collapse of the cervical veins, the so-called Friedreich's sign, is not of much moment.

*Palpation.*—The apex beat is fixed, and turning the patient on the left side does not alter its position. On placing the hand over the heart a diastolic shock or rebound is felt, which some regard as the most reliable of all signs of adherent pericardium.

*Percussion.*—The area of cardiac dulness is usually much increased. In a majority of instances there are adhesions between the pleura and pericardium, and the limit of cardiac dulness above and to the left may be fixed and is uninfluenced by deep inspiration. This is an uncertain sign, as there may be close adhesions between the pleura and pericardium and between the pleura and chest wall, which allow a considerable degree of mobility to the edge of the lung.

*Auscultation.*—The phenomena are variable and uncertain. In children, following rheumatic fever, endocarditis is usually present. Even in the absence of endocarditis, when the dilatation reaches a certain grade, there are murmurs of relative insufficiency, which may be present not only at the mitral but also at the tricuspid and pulmonary orifices. Theodore Fisher pointed out that there may be a well-marked presystolic murmur with adherent pericardium. Occasionally the layers of the pericardium are united in places by strong fibrous bands, 5-7 mm. long by 3-5 mm. wide. In one such case Drasche heard a remarkable whirring, systolic murmur with a twanging quality.

The pulsus paradoxus, in which during inspiration the pulse-wave is small and feeble, is sometimes present, but it is not a diagnostic sign of either simple pericardial adhesion or of the cicatricial mediastino-pericarditis.

The *prognosis* depends largely on the extent of the adhesions but especially on the state of the myocardium. Adhesions sufficient to give marked signs usually mean a seriously impaired heart. *Treatment* has to be directed to the heart muscle and is largely that of myocarditis. *Cardiolysis*, Brauer's operation, has been helpful in a few cases. Four or five centimetres of the fourth, fifth, and sixth left ribs with a couple of centimetres of the corresponding cartilages are resected, by which the heart's action is less embarrassed. It is a justifiable procedure in selected cases—in, for example, a child with a very large, tumultuously acting heart, with much bulging of the chest.

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### II. OTHER AFFECTIONS OF THE PERICARDIUM

**Hydropericardium.**—The pericardial sac contains post mortem a few cubic centimetres of clear, citron colored fluid. In connection with general dropsy, due to kidney or heart disease, more commonly the former, the effusion may be excessive, adding to the embarrassment of the heart and the lungs, particularly when the pleural cavities are the seat of similar transudation. There are rare instances in which effusion into the pericardium occurs after scarlet fever with few, if any, other dropsical symptoms. Hydropericardium is frequently overlooked. The features are much as in pericarditis with effusion except for the absence of pericardial friction. The treatment is that of the original condition with tapping if the symptoms demand it.

In rare cases the serum has a milky character—chylopericardium.

**Hæmopericardium.**—This is met with in aneurism of the first part of the aorta, of the cardiac wall, or of the coronary arteries, and in rupture and wounds of the heart. Death usually follows before there is time for the production of symptoms other than those of rapid collapse and heart failure. In rupture of the heart the patient may live for many hours or even days with symptoms of progressive heart failure, dyspnoea, and the signs of effusion. In the pericarditis of tuberculosis, cancer and nephritis, and in old people the exudate is often blood stained.

**Pneumopericardium.**—This is an excessively rare condition, of which Walter James was able to collect only 38 cases in 1903. We have met with but one instance, from rupture of a cancer of the stomach. Perforation of the sac occurred in all but 5, in which the gas bacillus was the possible cause, as in Nicholl's case in which this organism was isolated. Seven cases were due to perforation of the œsophagus and eight to penetrating wounds from without. The physical signs are most characteristic. A tympany replaces the normal pericardial flatness. On auscultation there is a splashing, gurgling, churning sound, called by the French *bruit de moulin*. This was described in 19 of the cases collected by James. Of the 38 cases, 26 died.

**Calcified Pericardium.**—This remarkable condition may follow pericarditis, particularly the suppurative and tuberculous forms; occasionally it extends from the calcified valves. It may be partial or complete. Of 59 cases collected by A. E. Jones, in 38 there were no cardiac symptoms. Case, in reporting an instance, found 90 recorded cases (1923). It is usually found at autopsy. There may be the signs of adherent pericardium. The X-ray study may give the diagnosis by the dense or mottled shadow of the heart.

# SYMPTOMATIC AND MECHANICAL DISORDERS

## B. DISEASES OF THE HEART

### I. SYMPTOMATIC AND MECHANICAL DISORDERS

#### I. SYMPTOMATIC DISORDERS

**Introduction.**—There are a number of disturbances referred to the heart which cannot be termed diseases—the term symptom-complex is a better designation. They may occur without any sign of organic cardiac disease but frequently cause extreme distress to the individual. It is not possible to group them in any systematic way. In some there are only subjective sensations, in others these occur with objective findings. We should remember that back of subjective disorders there is some cause and the effort should always be made to find it. Disturbances in the nervous system and in the internal secretions, infection in some form, unrecognized myocardial disease and the effects of toxic agents are particularly important.

(1) **Heart Consciousness.**—In health we are unconscious of the action of the heart. A not infrequent indication of debility or overwork is the consciousness of the cardiac pulsations which may be regular or irregular. This may be most evident when the patient is lying down. It is usually due to nervous fatigue, some form of debility or anæmia, or flatulence. It may be present with organic disease and hypertrophy.

(2) **Cardiac Pain.**—This may be referred to the whole præcordia or to local areas, most often about the apex or outside it. The area corresponds to the distribution of the eighth cervical to the fourth dorsal segments. A distinction should be made between aortic pain (aortitis, acute and chronic, most cases of angina pectoris, and aneurism) and cardiac pain. The former is usually felt over the upper part of the sternum and may be referred to the arms. It is important to secure an exact statement of the seat of pain. The influence of exertion, emotion, fear and excitement in causation is important. Many serious forms of heart disease are unaccompanied by pain. There are many causes for more or less persistent cardiac pain: (1) Myocardial, in which the pain is prolonged and usually most marked at the left border of the heart. It may amount only to a feeling of deep-seated discomfort or pressure, or a sense of weight or oppression. (2) Dilatation. (3) Pericarditis. (4) Valvular disease, especially aortic. (5) Disease of the coronary arteries. (6) Certain toxic influences, especially tobacco. (7) With the "Effort Syndrome." (8) Angina pectoris (some cases). (9) With digestive disturbances, especially distention and heart-burn. (10) In a large group in which no evidence of cardiac disease can be found and often termed "cardiac neurosis," which means little. This is common in women, especially at the menopause, and is especially marked in those who are "neurotic." Two forms are common: in one there is a dull more or less continuous pain and in the other sharp stabbing pains of short duration. Emotion is a frequent exciting cause. In some a definite disturbance of sensation can be found, usually near the apex. The term "pseudo-angina pectoris" should be dropped from our terminology. It has no set meaning and is very variously employed. Some use it as synonymous with vaso-motor angina pectoris. The group includes cases

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neurotic persons or in those who have used too much tobacco. The attacks have no necessary relation to exertion and may come on at night or when the patient is at rest; they are commoner in women and may occur at any age; and are not associated with demonstrable organic disease of the heart or aorta. The attacks may last for an hour or longer. There is often a feeling of distention and the patients are usually restless, making complaint of a number of symptoms. It must not be forgotten that there are cases of mild angina pectoris. It is safer to regard doubtful cases as examples of this than to label them "pseudo-angina."

The *diagnosis* of pain is based on the patient's statement; the estimation of its severity is made by observation; the recognition of its cause demands thorough study. Careful search should be made for organic disease; always suspect this until its absence is proved. Particular attention should be given to the state of the nervous system. The source of pain mistakenly regarded as cardiac but due to disease elsewhere is usually recognized by a thorough examination.

The *treatment* must be based on accurate diagnosis. In the "nervous group," the meaning of the symptom should be explained and every effort made to correct the causal factors. The use of bromides is indicated until there is improvement in the general condition. A dose of aromatic spirit of ammonia with valerian or Hoffman's anodyne is often helpful in an attack.

(3) **"Effort Syndrome," "Neuro-Circulatory Asthenia," "Disordered Action of the Heart," "Irritable Heart."**—The condition to which these terms are applied does not represent a specific disease but a combination of symptoms in which shortness of breath, fatigue, and vaso-motor disturbances are the principal features. The condition is not confined to soldiers; it occurs in civil life, and in females and children as well as in men. The subjects are usually of a sub-normal type physically and unable to do heavy physical work. The etiological factors are many. The general "make-up" is largely responsible. Infection may play a part, especially rheumatic fever, tonsillitis, influenza, focal infection, etc.; syphilis plays a very small part. Hyperthyroidism is a factor in a small percentage only. Disturbance of the central nervous system is important. Certain of the patients are of the visceroptotic build, with long thin bodies and poor muscular development, and cardioposis ("dropped heart") is relatively common. Stress and strain which they are unfitted to endure is a common determining factor in war.

*Symptoms.*—Shortness of breath, rarely at rest, but almost invariably on exertion, is the most frequent complaint, and is increased by effort, especially if hurried. With this are severe fatigue and exhaustion, sometimes with tremor. Pain is common, usually precordial or in the lower left costal region, and increased by exercise. Precordial tenderness and disturbance of sensation may accompany it. Palpitation of the heart on exertion and excitement often occurs. Syncope is not uncommon. Giddiness is frequent and may occur with change in position or on exertion. Vaso-motor phenomena are common; the hands and feet are blue, there is profuse sweating, and dermographia is marked. The patients show nervous instability, and are easily upset. The pulse rate is increased and responds quickly to exertion. The return to normal after exercise is slow. The blood pressure tends to be low. The heart shows an absence of signs of myocardial disease. Care must be

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taken to recognize the condition in which an overacting "nervous" heart simulates mitral stenosis.

In *treatment* any suggestion of "heart disease" should be avoided, and every effort made to explain the condition. Search should be made for the etiology and a causal factor treated if possible, especially a focus of infection. Tobacco and alcohol should be strictly forbidden. The whole method of life should be reviewed and every effort made to improve the general health by proper exercise, bathing and good hygiene. Cardiac drugs are not needed, but general tonics should be given if indicated.

(4) **Cardiac Asthenia in Children.**—This may occur at any age in children but especially about puberty. The complaints are of dizziness, weakness, discomfort about the heart and sometimes of syncope. The children are often pale and flabby with little endurance, and show vaso-motor instability. The heart rate is increased and there may be sinus irregularity or extra-systoles. The heart may be centrally placed and occasionally is slightly enlarged. A systolic murmur at the apex is common. Care should be taken to exclude myocardial disease or a focus of infection. These children should have long hours of rest and take regular exercise, carefully supervised and increased gradually. Cool sponges should be given if possible. Anæmia should be treated.

(5) **Palpitation.**—In health we are unconscious of the action of the heart. One indication of cardiac failure or stress is the consciousness of the cardiac pulsations, which may, however, be perfectly regular and orderly. This is not palpitation. The term is properly limited to irregular, rapid, or forcible action of the heart perceptible to the individual. Extra-systoles, auricular fibrillation and paroxysmal tachycardia are present in some cases.

*Etiology.*—The expression "perceptible to the individual" covers the essential element in palpitation of the heart. The most extreme disturbance of rhythm may be unattended with subjective sensations or distress, and there may be no consciousness of disturbed action. On the other hand, there are cases in which complaint is made of distressing palpitation and sensations of throbbing, in which examination reveals a regular heart, the sensations being entirely subjective. This occurs in a large group of cases in which there is increased excitability of the nervous system. Palpitation may be a marked feature at puberty, at the climacteric, and occasionally during menstruation. It is common in anæmia, hysteria and neurasthenia, and particularly with dyspepsia. Emotions, such as fright, are common causes of palpitation. It may occur as a sequence of the acute fevers. Females are more liable to the affection than males.

In a second group the palpitation results from the action upon the heart of certain substances, such as tobacco, coffee, tea, and alcohol. And, lastly, palpitation may be associated with organic disease of the heart, either of the myocardium or valves. As a rule it is a purely nervous phenomenon, seldom associated with organic disease in which the most violent action and extreme irregularity may exist without a subjective element of consciousness of the disturbance. It occurs frequently with hyperthyroidism.

*Symptoms.*—In the mildest form, such as occurs during a dyspeptic attack, there are slight fluttering of the heart and a sense of what patients sometimes call "goneness." In more severe attacks the heart beats violently, its

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palpitations against the chest wall are visible, the rapidity of the action is much increased, the arteries throb forcibly, and there is a sense of great distress. In some instances the heart's action is not quickened. The most striking cases are in neurasthenic women, in whom the entrance of a person into the room may cause violent action of the heart and throbbing of the arteries. The pulse may be rapidly increased until it reaches 150 or 160. A diffuse flushing of the skin may appear at the same time. After such attacks there may be the passage of a large quantity of pale urine. In many cases of palpitation, particularly in young men, the condition is at once relieved by exertion.

The examination usually shows a heart of normal size. The sounds, the shock of which may be very palpable, are clear, ringing, and metallic, but not associated with murmurs. The second sound at the base may be accentuated. A murmur may be heard over the pulmonary artery or at the apex in cases of rapid action in neurasthenia or severe anaemia. The attacks may be transient, lasting only a few minutes, or may persist for an hour or more. In some instances any attempt at exertion renews the attack. Sometimes in vigorous young adults who are upset nervously, especially after exertion or during excitement, the signs of mitral stenosis are simulated. There is a systolic shock preceded by a suggestion of a thrill. On auscultation it may be difficult to decide whether or not there is a short presystolic murmur. A short period of observation usually removes the uncertainty and the administration of amyl nitrite, which increases the murmur of mitral stenosis, is an aid. Organic murmurs are sometimes increased by pressure on the eyeballs.

The *diagnosis* should always include the conditions which are responsible. Nervous states (especially the anxiety neuroses and those due to disturbance in the sexual sphere), anaemia, gastro-intestinal disorders, and particularly the possible influence of the thyroid gland should be considered. In the condition termed *phrenocardia* there are palpitation, pain in the cardiac region or to the left of the apex, and respiratory disorder shown by frequent attempts to take a deep breath. There may be spasm of the diaphragm with cardiop-tosis. The *prognosis* is that of the underlying condition, which it may be difficult to remedy.

*Treatment.*—An important element is to get the patient's mind quieted and assure him that there is no actual danger. The mental element is often very strong. If an underlying cause can be found this should receive attention. Before using drugs, it is well to try the effect of hygienic measures. As a rule, moderate exercise may be taken with advantage. Regular hours should be kept, and at least ten hours of the twenty-four should be spent in the recumbent posture. A tepid bath may be taken in the morning, or, if the patient is weak and nervous, in the evening, followed by a thorough rubbing. Hot baths and the Turkish bath should be avoided. The dietetic management is important and it is best to prohibit alcohol, tea, and coffee absolutely. The diet should be light and the patient should avoid taking large meals. Articles of food known to cause flatulency should not be used. If a smoker, the patient should give up tobacco. Sexual excitement is particularly pernicious, and the patient should be warned especially on this point. The cases of palpitation due to excesses or to errors in diet and dyspepsia are readily remedied by hygienic measures.

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A course of iron is often useful. Strychnia is particularly valuable, and is best administered as the tincture of *nux vomica* in large doses. Very little good is obtained from small quantities and it should be given freely, 20 minims (1.3 c. c.) three times a day. If there is great rapidity of action, aconite may be tried. There are patients with sleeplessness and restlessness who are greatly benefited by the bromides. *Digitalis* is very rarely indicated, but in obstinate cases it may be tried with the *nux vomica*.

### II. MECHANICAL DISORDERS OF THE HEART-BEAT

**Normal Mechanism.**—The normal heart-beat is initiated by a stimulus which originates in a small mass of highly specialized tissue situated at the junction of the superior vena cava and the right auricle (sino-auricular node). This node under normal conditions determines both the rate and rhythm of the heart, and is, therefore, designated the "pacemaker." The excitation wave, immediately followed by muscular contraction, first sweeps over the walls of the auricles and then passes to the ventricles by a special conduction system, having its beginning in the node of Tawara, situated low down on the right side of the inter-auricular septum, and continued forward and downward as a narrow neuro-muscular band (bundle of His), which upon reaching the intraventricular septum primarily divides into a right and left branch supplying the respective ventricles; after further multiple subdivisions, the end branches terminate as the fibres of Purkinje, bringing the conduction system into intimate anatomical and functional relation with the ventricular musculature. The mechanical and functional efficiency of the heart depends in considerable degree upon the integrity of these various structures, and the orderly exercise of their functions. In health, under ordinary circumstances, stimuli are rhythmically generated in the sino-auricular node at the rate of 72 per minute and reach the ventricular walls after a lapse of approximately 0.2 second, thereby insuring a sequential contraction of auricles and ventricles as a result of the stimulus wave. An epitome of the myocardial functions is as follows: "The muscular fibres of the heart possesses the power of rhythmically creating a stimulus, of being able to receive a stimulus, of responding to a stimulus by contracting, of conveying the stimulus from muscle fibre to muscle fibre, and of maintaining a certain ill-defined condition called tone" (Gaskell). Disturbance, impairment or loss of these functions occasion mechanical disorders of the heart-beat.

The stimulus and contraction normally proceed in this regular fashion but if the excitability of any other part is sufficiently increased, a stimulus may originate there, giving rise to irregular contractions. If the ventricle is the most excitable part, the stimuli arising there may pass to the auricle, causing a reversed rhythm. After contraction has occurred, for a time the muscle can not be stimulated to contract again (refractory period). The muscle either contracts with its full power or does not contract at all in response to stimulation, "all or none." The longer the interval since the previous contraction, the weaker the stimulus which produces contraction. Also the longer the period of rest (diastole) the better prepared the muscle is to contract. Increase in rate is largely at the expense of diastole, so that with rapid rates the rest period of the heart is markedly shortened. The

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Normal times are auricular systole .1 second, ventricular systole .3 second, and diastole .4 second. While the heart has the inherent power of originating stimuli and contracting, yet nervous control plays a part. This can influence all the functions of the heart muscle. The vagus depresses excitability, contractility, conductivity and tonic. Inhibition of the heart may result from excessive stimulus of the vagus. The sympathetic supply increases contractility and conductivity. Much of the importance of the heart action pertains to systole and diastole is less regarded. It might be well to give more attention to diastole, which is the rest period and which is specially associated with tonic, in the maintenance of which the vagus influences probably play a large part.

### A. DISTURBANCES OF RATE

(1) **Tachycardia** (with Normal Mechanism).—The rapid action may be perfectly natural. There are individuals whose normal heart rate is 100 or even more per minute. Emotional causes, violent exercise, and fevers all produce great increase in the rapidity of the heart's action. The extremely rapid action which follows fright may persist for days or even weeks. Cases are not uncommon at the menopause.

There are cases in which it depends upon definite changes in the pneumogastrics or in the medulla. Cases have been reported in which tumor or clot in or about the medulla or pressure upon the vagi has been associated with rapid heart. Tachycardia occurs under many conditions, such as hyperthyroidism, mitral stenosis (apart from fibrillation), interference with the vagus (mediastinal tumor, etc.), post-febrile conditions, anæmia, the effect of certain drugs (belladonna, thyroid extract), nervous disturbance, toxic states (tobacco), etc. The tachycardia may persist for months or indefinitely, and may interfere with the amount of exertion such persons can make; in addition there is a sense of weakness and sometimes syncope. The diagnosis of the cause is essential and on this the treatment must be based.

(2) **Bradycardia** (True).—Slow action of the heart is sometimes normal and may be a family peculiarity. Napoleon is stated to have had a pulse of only 40 per minute. In any case of slow pulse rate it is important first to make sure that the number of heart and arterial beats correspond. In many instances this is not the case, and with a radial pulse of 40 the cardiac pulsations may be 80, half the beats not reaching the wrist. The heart contractions, not the pulse waves, should be taken into account.

**Physiological Bradycardia.**—As age advances the pulse rate becomes slow. In the puerperal state the pulse may beat from 44 to 60 per minute, or may even be as low as 34. It is seen in premature labor as well as at term but the explanation is not clear. Slowness of the pulse is associated with hunger. Bradycardia depending on individual peculiarity is extremely rare.

**Pathological bradycardia** is met with under the following conditions: (a) In convalescence from acute fevers. This is extremely common, particularly after pneumonia, typhoid fever, influenza, and diphtheria. It is most frequent in young persons and in cases which have run a normal course. (b) In diseases of the digestive system, such as chronic dyspepsia, ulcer or cancer of the stomach, and jaundice. (c) In diseases of the respiratory system.

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Here it is by no means so common, but is not infrequent in emphysema. In diseases of the circulatory system. Bradycardia is not common in diseases of the valves. It is most frequent in fatty and fibroid changes in the heart, but is not constant in them. Sino-auricular block is a rare cause. (e) It occurs occasionally in nephritis and may be a feature of uræmia. (f) From the action of toxic agents. It occurs in uræmia, poisoning by lead, alcohol, and follows the use of tobacco, coffee, and digitalis. (g) In constitutional disorders, such as anæmia and diabetes. (h) In diseases of the nervous system. Apoplexy, epilepsy, one stage of tuberculous meningitis, cerebral tumors, affections of the medulla, and diseases and injuries of the cervical cord may be associated with a slow pulse. In general paresis, dementia præcox, mania, and melancholia it is not infrequent. (i) It occurs occasionally in affections of the skin and sexual organs, and in sunstroke, or in prolonged exhaustion from any cause.

*Treatment.*—For the bradycardia itself little can be done. The cause should receive attention.

### B. DISTURBANCES OF RHYTHM AND FORCE

1. **Sinus Arrhythmia.**—This depends on changes in the control of the sino-auricular node in which the effect of vagus influence is important. It is frequently seen in connection with respiration, especially in deep breathing. The rate increases with inspiration and slows with expiration. This is common in young children and about the time of puberty, and is seen occasionally in adults. In some cases it may be responsible for attacks of faintness or syncope, sometimes with a slow rate and a low blood pressure. The occurrence

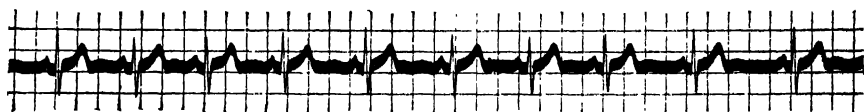


FIG. 1.—SINUS ARRHYTHMIA.

This exhibits a variation occasioned by a difference in length of diastoles. The systoles are the product of a normal mechanism and are unevenly spaced but show no variation.

of irregularity, also with slow pulse rate, and which has no order in its occurrence, is sometimes seen. This may occur after the administration of digitalis, in rheumatic myocarditis or with the bradycardia so common after pneumonia. The condition is not serious in any way.

*Diagnosis.*—This is usually clear. The irregularity is of the whole beat and the pulse and apex beat correspond. The occurrence with respiration is significant. Exercise, fever and atropine usually abolish this irregularity.

*Treatment.*—None is required and this condition should not be regarded as an indication for rest or lessened activity.

2. **Extra-systole (Premature Contraction).**—This is a common form of irregularity, to understand which it must be remembered that to a stimulus strong enough to set up a contraction the heart answers with all the contractility of which it is capable at the moment (Bowditch's law of maximal contraction). A second property of the heart muscle is that it pos-

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causes a "refractory phase" in which normally it is not excitable, or answers only to very strong stimuli. The auricle, the auricular-ventricular node or the ventricle may be more excitable than the sinus and originate an abnormal stimulus for contraction, the sinus rhythm not being altered. Extra-systoles may occur regularly or irregularly and often or seldom. Auricular and ventricular extra-systoles are more common than the nodal form. (a) *Auricular extra-systole*. The premature stimulus and contraction of the auricle usually cause a premature contraction of the ventricle, but the stimulus may travel slowly, so that the auricular contraction interval is prolonged, or it may not reach the ventricle—it is blocked. (b) *Nodal or auricular-ventricular extra-systole*. The stimulus arises in the auricular-ventricular junctional tissue and passes both to the auricle and ventricle. The two may contract together or either may be first.

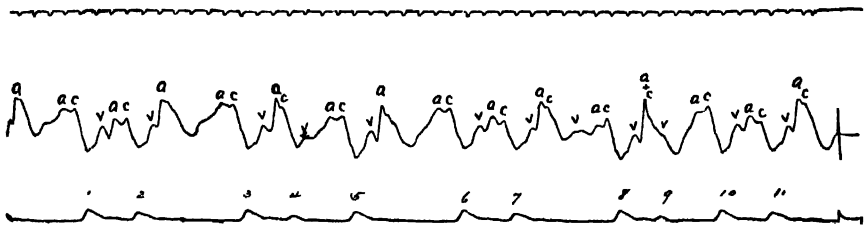


FIG. 2.—FREQUENT PREMATURE CONTRACTIONS.

The radial pulse waves numbered 1, 2, 3, 5, 6, 7, 8, 10, and 11 are the result of a normal mechanism; those numbered 4 and 9 are premature; incomplete premature ventricular contraction occasioning pulse intermission occurred after the 2, 5, 7, and 11 waves. The time occupied by a normal beat and an intermission is exactly equal to two normal pulse periods. There is a demonstrable tendency to rhythm despite the marked grade of irregularity. The venous pulse (upper curve) shows the auricular contractions (a waves) occurring regularly. The normal relation of the ventricular waves (c and v) to the auricular wave is deranged at the period of disturbance. The irregularity is due to premature contraction of the ventricle occurring in advance of the arrival of the normal excitation wave from the auricle.

(c) *Ventricular extra-systole*. The extra impulse arising in the ventricle and causing it to contract anticipates the next regular impulse from the auricle which arrives when the ventricle is in the "refractory phase" and hence it does not contract, so that the auricular impulse is wasted. The interval which follows the premature contraction will therefore be longer than normal (compensatory pause). In consequence of this prolonged interval of rest the next contraction is likely to be more forcible than normal. If the ventricular extra-systole occurs early enough to permit the next regular auricular stimulus to arrive when the ventricle can contract, there is a premature contraction between two normal beats. The premature contraction is not as strong as a normal one and hence the pulse wave resulting is smaller. If the contraction is very feeble (and the earlier it occurs in diastole the weaker it is) the aortic valves may not be opened or the wave may not reach the wrist. This results in a dropped beat. If an extra-systole occurs regularly after each normal beat, then the apex impulse and the pulse wave occur in pairs—*pulsus bigeminus*; or if the extra-systole occurs after two normal beats, the impulse and pulse occur in threes—*pulsus trigeminus*, and so on.

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On auscultation two sounds are heard with the extra-systole, if the aortic valves are opened, otherwise only a first sound. Evidently there can be many variations in the sounds and character of the pulse. Graphic records are usually necessary to distinguish between the auricular, nodal and ventricular forms of premature contractions. If a murmur is present it may be absent or less loudly heard with the premature beat. Fever, exercise, a change in posture, and a rapid heart rate may cause the temporary disappearance of extra-systoles. They tend to be more frequent with a slowing heart, as after exercise, on taking the recumbent posture or after fever.

*Symptoms.*—The irregularity, inequality, and intermission of the pulse met with in every day experience are largely due to extra-systoles, which may present all sorts of combinations and groupings, depending upon whether the extra pulse beats are perceptible or not. There may be no actual pathological change, and so far as the maintenance of the circulation is concerned the heart may be acting in a satisfactory manner. The subjective sensations



FIG. 3.—PREMATURE CONTRACTIONS OF VENTRICULAR ORIGIN.

The grouping of pulse beats shows the manner of production of bigeminal, trigeminal, and quadrigeminal pulses.

vary greatly. In some the extra-systoles are not noticed but many complain of a variety of symptoms and especially of the pause with the succeeding strong contraction. Some patients are greatly disturbed by them.

Extra-systoles occur at all ages and under the most varied conditions but are most common in persons over fifty. There are several classes of cases. The irregularity may be a life-long condition, without any recognizable disease and without any impairment of the function of the heart. This may be a peculiarity of the heart-muscle of the individual, who has extra-systole for the same reason—physiological but not well understood—as the dog and horse, in which animals it is common. The late Chancellor Ferrier, of McGill University, who died at the age of eighty-seven, had an extremely irregular heart action for the last fifty years of his life. In debilitated and neurasthenic persons there may be an irritable weakness of the heart associated with extra-systole and palpitation of a distressing character. In a second group toxic agents, as tobacco, tea, coffee, or the toxins of the infectious diseases are responsible. Digitalis may be a cause. Even reflexly, as in flatulent dyspepsia, extra-systoles may arise. Thirdly, a high blood pressure can set up extra-systoles; also change in posture. And, lastly, organic disease of the heart itself, especially myocardial.

The *significance* of premature contractions is not always easy to determine. They are often temporary, especially in young persons, but should not be regarded lightly. It is wiser to regard them as meaning some pathological

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change until the contrary is proved than to make light of them and recognize the error later. In those who have reached fifty years of age they may be the warning of serious myocardial damage. The patient seen to-day with extra-systoles may return with auricular fibrillation in two or three years. The *diagnosis* is usually easy. The occurrence of a premature contraction is felt at the apex and wrist and heard on auscultation. In the cases in which the premature contractions do not send a pulse wave to the wrist, the impulse may be felt at the apex or the sound heard. Tracings are rarely necessary.

*Treatment.*—This must depend on the other conditions found and not on the extra-systoles themselves. The nature of the process should be explained. In nervous patients, bromide is indicated. Quinine sulphate (gr. v-x, 0.3-0.6 gm., three times a day) with strychnine (gr. 1/30, 0.002 gm.) sometimes seems to lessen or stop extra-systoles. The condition itself does not require digitalis.

(3) **Simple Paroxysmal Tachycardia.**—This is characterized by paroxysmal attacks, beginning and ending abruptly, in which the heart rate increases to between 100 and 200 a minute (the common rate is between 140 and 190). The abnormal impulses arise from a new focus which may be in the auricle or ventricle, usually in the auricle. They represent "essentially a regular series of extra-systoles" (Lewis).

It may occur at any age but is most frequent in young adults, and more often in males. There may be definite cardiac disease but some patients show no sign of any lesion in the intervals. Naturally one is suspicious of some underlying factor (myocardial). Exertion, emotion or digestive disturbance may initiate an attack but in some cases no cause can be given. The duration of an attack varies from a few seconds to ten or more days.

The *symptoms* vary greatly with the duration and severity of the attack. A striking feature is the abrupt onset. In the very short attacks the patient may not be conscious of any disturbance or make any complaint. In more marked attacks there may be discomfort and palpitation, with weakness, sweating and gastric disturbance. Thoracic pain of varying distribution is common, sometimes with disturbance of sensation. If dilatation of the heart follows there are the symptoms associated with it. In the examination there may be little except the rapid heart and the general condition is often good. There may be marked pulsation in the veins of the neck. The heart rate should be determined by auscultation. The sounds are very short and sharp, like the fetal heart sounds. If there has been a previous murmur it may have disappeared. Enlargement of the heart, passive congestion of the lungs, sometimes with bloody sputum, cyanosis, œdema, and enlargement of the liver with abdominal tenderness may be found. These disappear rapidly when normal rhythm is restored.

In *diagnosis* the history of previous attacks with an abrupt onset and a sudden termination is important. The cases of tachycardia of other etiology rarely cause doubt. The rapid rate with loss of compensation should not cause difficulty. Change in posture and exertion do not alter the rate in paroxysmal tachycardia. In cases of doubt a tracing is diagnostic.

The *outlook* is good but always has an element of uncertainty. In prolonged attacks with marked disturbance of the circulation there is always some danger. The condition of the heart between attacks and the behavior of the muscle during the attack are important points. As to the patient becoming

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free of the attacks, it is difficult to speak with any certainty. The condition is compatible with long life. The late H. C. Wood had a patient aged eighty-seven, who had attacks at intervals for fifty years in which the pulse rate was usually 200. The taking of ice water or strong coffee arrested the attacks.

**Treatment.**—In an attack the patient should be quiet and in the position which gives him the greatest comfort. The diet should be liquid. If there is gastric disturbance, the giving of sedatives and alkalis may be useful. An ice bag applied over the præcordia often gives relief, if it does not stop the attack. The most diverse procedures may stop an attack, such as placing the head between the knees, being suspended with the head down, pressure on the vagus in the neck, or on the eye-balls, any sustained respiratory effort, the production of vomiting, the application of a tight abdominal binder, etc. The giving of strophanthin (gr. 1/250, 0.0026 gm.), quinine dihydrochloride (gr. v, 0.3 gm.) or epinephrine, (m x, 0.6 c. c. of a 1-10,000 solution) intravenously may be effectual. Chloral hydrate or morphia may be given to secure sleep. Any indicated symptomatic treatment should be given. Between attacks, any exciting cause should be avoided, the general health improved if possible, and attention paid to any gastro-intestinal disturbance. The wearing of an abdominal binder is sometimes useful.

(4) **Auricular Flutter.**—In this rather rare condition the impulses arise in the auricle, probably from a circular wave of contraction, which cause it to beat rhythmically at a rate of 200 to 350 per minute. As Lewis says, this may not be readily distinguished from paroxysmal tachycardia but when the rate is over 200 special characteristics appear. Lewis regards the circus movement as important in auricular flutter. The difference between it and fibrillation is largely one of degree. Heart-block is almost always present with it, the ventricular rate being half that of the auricle; 2:1 block is common but other ratios occur. There may be a change from one ratio to another. The rate of the auricle is regular; the ventricle is usually regular but sometimes irregular. It is most frequent in advanced years, more common in males and usually associated with arterio-sclerosis and myocarditis but may occur with acute infections or after operation.



FIG. 4.—AURICULAR FLUTTER.

The curve shows a series of regular waves due to auricular contractions, interrupted by sharp spikes due to ventricular contractions. The ratio between auricular contractions and ventricular responses varies (2:1, 3:1 and 4:1) and averages 3:1. The slower ventricular rate is caused by a partial heart-block. The auricular rate is 210 per minute; the ventricular responses average 70.

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The *symptoms* are fewer than might be expected and depend on the state of the myocardium. There may be a complaint of palpitation and attacks of syncope. Occasionally the ventricle takes the auricular rate, with which the condition is very grave, but such attacks are usually of short duration. The diagnosis may be possible only by electrocardiographic tracings if the rate of the ventricle is not very rapid; otherwise a rate of 130 or over is very suggestive. The duration is variable; it may last a very short time or persist for months. It may return to normal rhythm or pass into auricular fibrillation abruptly. The attacks begin and end abruptly. The effect on the heart depends greatly on the condition of the myocardium, but the duration and ventricular rate also have an influence. There is always a probability of a return of the condition. The *treatment* consists in the use of digitalis or strophanthus in full doses and until a definite effect is produced. As a rule the administration of digitalis should be continued in amounts sufficient to control the rate of the ventricle.

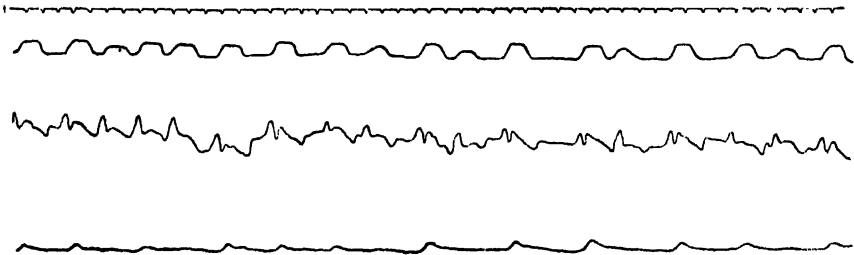


FIG. 5.—AURICULAR FIBRILLATION.

Several cardiac contractions at the apex (upper tracing) produced no pulsation at the wrist; others are so small as not to be felt. Synchronous counts at the apex and wrist for 10 seconds show 18 and 13 impulses respectively. The jugular shows only *c* and *v* waves, due to ventricular activity. Waves due to contraction of the auricles are absent, since they have ceased to act as efficient contracting chambers.

(5) **Auricular Fibrillation.**—This common form of cardiac irregularity is exceedingly important to recognize clinically. It is seen most frequently in the last stages of mitral stenosis, in which the pulse shows extreme irregularity, which, when once established, is usually permanent. Transient and recurring forms are observed as a result of acute conditions and preceding the permanent form. A study of its features in this condition gave Mackenzie the clue to its explanation. He found that in certain cases the transition from regular to irregular pulse of this type occurred with suddenness, and that, whereas before the irregularity supervened the jugular pulse showed the normal features in the presence of auricular carotid and ventricular waves, with a marked presystolic murmur and thrill at the apex, after the irregularity was established, the auricular wave disappeared from the jugular pulse and the presystolic murmur from the apex. The inference drawn was that the right auricle was so dilated as to prevent the formation of a normal auricular contraction. Complete proof of the cause of this condition was supplied by Lewis, who found that patients with this irregularity showed in galvanometric tracings from the auricle numerous small and continuous waves, similar to those obtained in the dog after fibrillation of the auricle has been

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induced by faradic stimulation of the appendix of the right auricle, or by ligation of the right coronary artery. The auricles do not contract normally but are in diastole with many fibrillary twitchings. The numerous abnormal impulses come to the auricular-ventricular bundle but only some of them are able to pass and these reach the ventricle in an irregular fashion. Hence the contractions of the ventricle are disturbed and irregular. The state of the bundle determines how many impulses pass and hence the ventricular rate shows great variation. Heart-block and auricular fibrillation may occur together. Lewis regards auricular fibrillation as due to a *circus movement* in the auricle, "a single wave is propagated and revolves perpetually upon a re-entrant path." This is repeated as an average 450 times per minute and controls the beating of the auricle. The relation of the refractory period to the speed at which the wave moves is important. If the advancing wave of contraction reaches muscle in the refractory period, evidently the con-



FIG. 6.—AURICULAR FIBRILLATION.

The irregularity of the pulse is evident, and is further shown by accurate measurement of the duration of each cycle. The beats averaged 65. The low rate was occasioned by associated sclerosis of the bundle limiting the number of stimuli reaching the ventricles from the auricles.

traction will stop. Quinidine acts by lengthening the refractory period so that the head of the circulating wave catches up with its tail, so to speak. When this happens the circus movement ceases. But quinidine also slows conduction which favors the circus movement. One effect tends to shorten the gap between "the crest of the movement and its wake," the other to lengthen it. Only if the first predominates will the abnormal movement terminate.

Auricular fibrillation forms a large proportion of the cases showing myocardial failure with an irregular pulse. Many of the contractions fail to give a pulse wave at the wrist (pulse deficit). Of etiological factors the most important is mitral stenosis. It is essentially a sign of marked myocardial disease. It occurs occasionally in acute infectious diseases. The average age of onset in those with a previous history of rheumatism is 30 to 40; in the non-rheumatic group it is between 50 and 60.

The symptoms depend largely on the associated conditions and are those of marked myocardial failure. The ventricular rate has some influence, as when it is very rapid (120-160), the distress and general symptoms of dilatation are more marked. The pulse is extremely irregular in every way and an irregular pulse with a rate over 120 is usually due to fibrillation. The more rapid the rate, the greater the irregularity. The diagnosis is clear with a

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very rapid heart, as all irregular hearts with a rate of 120 or more have auricular fibrillation, but when the rate is below 100 there may be slight difficulty until a careful study is made. The marked irregularity in every way—force, rhythm and rate—is usually sufficient. Tracings remove any difficulty.

In *prognosis* the occurrence of fibrillation is of grave omen. The condition is compatible with life for years but means serious myocardial damage. The ventricular rate is of value, a persistent rate of 120 or over means a grave outlook and each increase in rate above this is more serious. The influence of treatment is of value in estimating the outlook.

*Treatment.*—For the general condition of the heart the problem is that of myocardial insufficiency, and for this and for the fibrillation the remedy is digitalis, which acts by blocking the passage of many of the impulses from auricle to ventricle. The dosage is that which keeps the heart at the best possible rate, and must be decided for each patient. It depends somewhat on the severity of the condition; the present tendency is to give larger doses than formerly. In any case the object is to produce the desired effect, whatever dose is required. Most of the patients should continue the use of digitalis permanently.

The use of *quinidine* has been successful in a considerable number of cases. It probably depresses conduction. As a rule the patients should first have the advantage of digitalis, but this should be stopped for several days before quinidine is given. The patients should be carefully chosen; those in whom the fibrillation is of recent development (within six months); those without marked evidence of myocardial degeneration, or arterio-sclerosis, are the most favorable. In patients with cardiac disease of long duration with decompensation, especially if the liver is enlarged, acute or subacute endocarditis, multiple valve lesions, coronary artery disease, heart-block or angina pectoris, quinidine is not likely to be of use. It is well to give two doses of 3 grains (0.2 gm.) on the first day. The next day it is given in doses of 6 grains (0.4 gm.) three times and continued for three or four days. If there is an effect the drug should be given in smaller doses (0.2 gm. once or twice a day) for some time. The duration of this has to be settled for each patient. Some prefer to give larger doses, especially if the usual dosage is without effect. The question of danger is unsettled; apparently the risk of embolism is comparatively slight. Various disturbances may occur, such as dizziness, weakness, palpitation, dyspnoea, diarrhoea, etc. Sudden collapse with unconsciousness or additional disturbance of rhythm may result. The drug should be stopped if any of these appear. If quinidine has an influence on the fibrillation, it is of value in determining how much part fibrillation plays in the myocardial failure.

(6) **Heart-block** (Stokes-Adams Syndrome).—In the adult heart the auriculo-ventricular bundle of His is 18 mm. long, 2.5 mm. broad, and 1.5 mm. thick; it arises in the septum of the auricles below the foramen ovale and passes downward and forward through the trigonum fibrosum of auriculo-ventricular junction, where it comes into close relation with the mesial leaflet of the tricuspid valve. Passing along the upper edge of the muscular septum, just where it joins with the posterior edge of the membranous septum, it radiates throughout the ventricles. If the function of the auriculo-

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ventricular bundle is impaired there may be a delay in the conduction of the impulse or it may be blocked completely. This may occur only with certain impulses (*partial heart-block*) or with all (*complete heart-block*). Intermittent and variable forms have been observed. In complete block the ventricles, released from the control of the normal pace-maker, assume their own rhythm (usually about 30 a minute).

**Etiology.**—Heart-block may occur at any age depending on the cause. It is more common in males. It is not infrequent, but usually temporary, in infectious diseases, especially rheumatic fever, influenza, diphtheria and pneumonia, but occurs in many others. In this group acute inflammatory changes or the influence of toxins may be responsible. Syphilis is an important cause owing

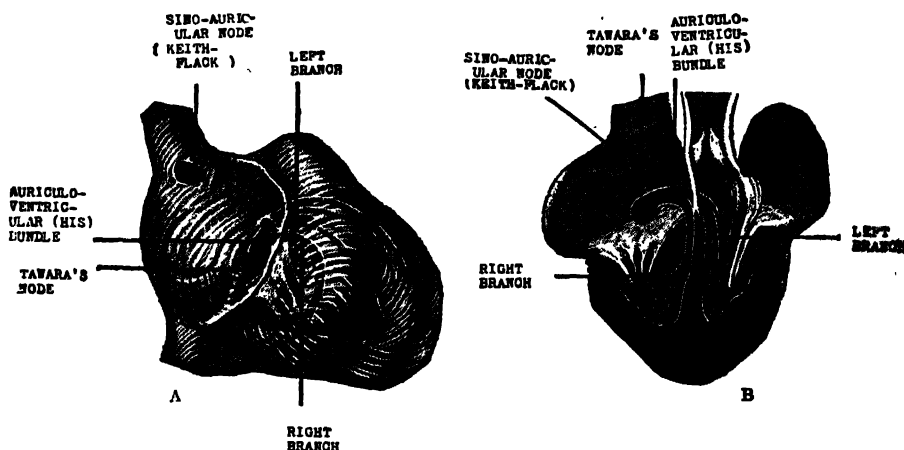


FIG. 7.—DIAGRAM SHOWING THE SINO-AURICULAR NODE AND THE AURICULAR BUNDLE. A, viewed from the right; B, cross section of the heart, viewed from the front. (Kindness of A. D. Hirschfelder.)

to the auriculo-ventricular bundle being affected in the myocardial involvement or by a gumma. Any form of myocarditis, acute or chronic, may be responsible. The action of digitalis in auricular fibrillation depends largely on its action on the impulses passing from auricle to ventricle and hence it is one of the causes of heart-block. The lesion in the bundle may be acute, usually in infections, or chronic, with fibrosis, gumma, etc. Heart-block usually occurs with marked myocardial degeneration, involving both the muscle and the conduction system.

The *symptoms* are variable, and depend to a considerable extent on the associated conditions. Some patients make little complaint, but dizziness, weakness and fainting attacks are not uncommon. In the more severe forms, the syncopal attacks are more frequent and severe. One variety is described under the Stokes-Adams syndrome. (It may be emphasized that this and heart-block are not synonymous terms.) Heart-block occurs without the Stokes-Adams syndrome and the syndrome may be due to other causes. The *signs* vary with the grade of block. An early manifestation may be reduplication of the first or second sound due to lengthening of the A-V interval which represents a delay in conduction. A dropped beat is easily recognized and if the ventricle is beating regularly at half the rate of the auricle (2:1 block)

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the pulse may be 40 to 50 a minute. Halving of the ventricular rate under digitalis therapy is always suggestive. It may be that the auricular rate can be counted by the pulsations in the veins of the neck. In *complete* block the ventricle beats at a rate below 35, and independently of the auricle.

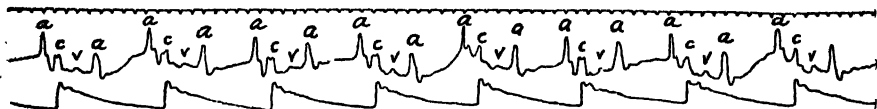


FIG. 8.—PARTIAL HEART-BLOCK WITH 2:1 RATIO.

The auricular rhythm indicated by the *a* waves in the venous pulse is regular; the ventricles respond regularly to every other auricular contraction. Auricular rate 66, ventricular and pulse rate 33.

While the diagnosis can often be made from the physical signs, tracings render it certain. The more frequent contractions of the auricle may be seen with the fluoroscope.

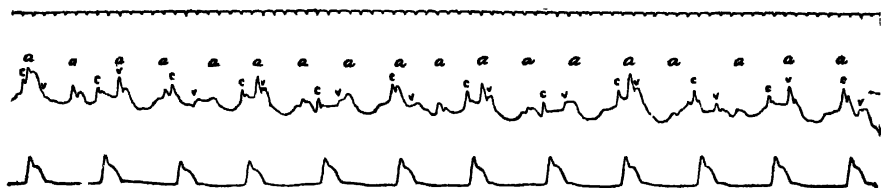


FIG. 9.—COMPLETE HEART-BLOCK.

The auricles and ventricles are pursuing independent, separate rhythms, indicated by the variable relation of auricular (*a*) waves to those resulting from the ventricular contraction (*c* and *v* waves). In the period of 16 seconds shown nearly every possible arrangement of these waves occurs. The auricular rate is 62.5; ventricular, 45.

*Stokes-Adams Syndrome.*—This may occur with heart-block, more often partial than complete, or in conditions in which the rate is low from other causes. Clinically this presents three features: (a) Slow pulse, usually permanent, but sometimes paroxysmal, falling to 40, 20, or even 6 per minute; (b) cerebral attacks—vertigo of a transient character, syncope, pseudo-apoplectiform attacks or epileptiform seizures; (c) visible auricular impulses in the veins of the neck, as noted by Stokes—the beats varying greatly; a 2:1 or 3:1 rhythm is the most common. There are several groups of cases. It is usually a senile manifestation associated with arterio-sclerosis. The cases in young adults and middle aged men are often myocardial and of syphilitic origin. There is a neurotic group in which all the features may be present, and in which post mortem no lesions have been found (Edes and Councilman). In some cases of slow pulse in this group the auricular as well as the ventricular rate may be slow and equal, the normal sequence of events being preserved; the origin of the condition is probably vagal. The outlook in this class of cases is good; in the others it is a serious disease and usually fatal, though it may last for many years. The cerebral attacks are due to anæmia of the brain or of the medulla in consequence of the imperfect ventricular action.

The *prognosis* in the cases of heart-block with acute infectious disease

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is usually good, with the exception of diphtheria and some cases of rheumatic fever. In the chronic forms the outlook is grave and sudden death is always possible. The ultimate outlook in cases of branch bundle block is very serious. The syncopal and convulsive attacks are always serious. In some of the cases due to syphilis proper treatment may result in great improvement. In

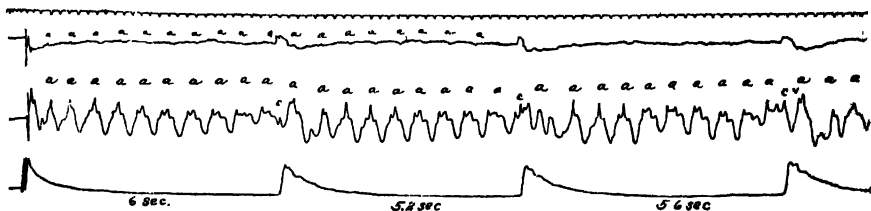


FIG. 10.—COMPLETE HEART-BLOCK WITH MARKED BRADYCARDIA.

Three contractions of the ventricles (upper curve) and three consequent radial pulse waves (lower curve) are recorded. The intervals elapsing between beats are shown. The ventricular contractions average slightly less than 11 per minute. The auricular contractions are indicated by the *a* waves in the venous pulse (middle curve) and show a regular rhythm and a rate of 110.

every case the state of the myocardium is important and with signs of its failure the outlook is grave.

**Treatment.**—If a cause, such as syphilis, is found, the indications are evident. Acute heart-block demands absolute rest and treatment directed to the general cardiac condition. Digitalis should be given with care. In partial block it may increase the difficulty and yet the heart muscle may be aided by it. In complete block it may be more useful and it cannot increase the block. The effect on the myocardium must decide. In partial heart-block the giving of atropine may be useful but rather in the cases due to acute infections than those with sclerotic processes. In the Stokes-Adams syndrome the injection of epinephrine (m 5-10, 0.3-0.6 c. c. of 1-1000 solution) subcutaneously (not intravenously) may have a marked effect which lasts for ten or twelve hours.

**Sino-Auricular Block.**—This is not common and may occur in healthy persons. Either the sino-auricular node does not function at intervals or the stimulus is not transmitted to the auricle. In consequence the auricle and ventricle fail to contract and there is a pause, usually less than the length of two cardiac cycles. There is a dropped impulse in the pulse. S. C. Smith found it in children after various infections and sometimes after exertion or excitement in apparently healthy persons. He regards it as probably not a pathological condition. Treatment is not required.

**Branch Bundle or Intraventricular Block.**—In this there is delay or block in the conductor system below the bifurcation of the bundle of His. The grade of block varies and the condition gives no distinctive signs. There is usually evidence of myocardial disease. The *diagnosis* can be made only by the electrocardiogram. It is of serious import. *Treatment* must be decided by the myocardial condition.

(7) **Alternation of the Heart.**—In this there is disturbance of the ventricular systole, so that larger and smaller amounts of blood are expelled by alternate contractions and consequently the pulse shows alternate large and small beats (pulsus alternans). It is seen in conditions of very rapid heart-rate, especially paroxysmal tachycardia and hyperthyroidism. Its occurrence

When the heart rate is normal or nearly so has a very different and more serious meaning. It is observed in a variety of conditions in which marked circulatory disease is present, in severe infections, especially pneumonia, in uræmia, in lead poisoning, and in patients under the influence of digitalis. It occurs most often in advanced life in males with myocardial degeneration. It is essentially due to a partial failure of contractility, the most important of the myocardial functions, and is associated with conditions which cause heart failure. It is comparatively common but frequently overlooked.

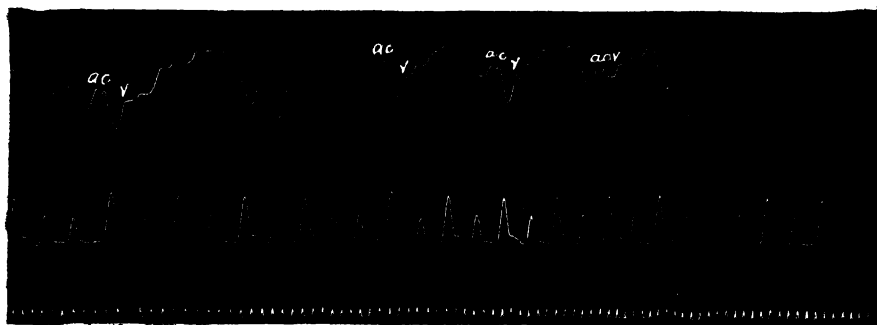


FIG. 11.—ALTERNATION OF THE PULSE.

The irregularity is of volume only, the sequence being regular. In two places the regular alternation of large and small beats is interrupted by the occurrence of two small beats.

The condition itself probably causes no *symptoms* but as it accompanies serious circulatory diseases, the features of these are present, such as dyspnoea, anginal pain, etc. It should be searched for in cases of hypertension, angina pectoris, myocarditis and when extra-systoles are present. It may be more evident after exertion, with the patient standing or after holding the breath. The variations may be felt by the finger occasionally but tracings give the most certain evidence. The difference in systolic pressure between the large and small beats may be an aid, as at one level only half the beats come through. Comparison with the heart rate distinguishes it from a dicrotic pulse. A bigeminal pulse due to extra-systoles may give difficulty but the regular intervals in pulsus alternans should decide. Excluding the cases of tachycardia and usually those due to digitalis, the *significance* of alternation is always serious. This applies particularly to the cases in which it is continuous, but in all it should be regarded as an evidence of great danger. Sudden death is comparatively common. The *treatment* is that of the underlying condition and special emphasis should be placed on rest, thorough and prolonged.

## II. AFFECTIONS OF THE MYOCARDIUM

### I. HYPERTROPHY

**Varieties.**—The heart enlarges to meet a demand for extra work, either general, as in the hypertrophy of work, or special to combat a deficiency of cardiac structure, such as a damaged valve. There are two forms, one in

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which the cavity or cavities are of normal size, and the other in which the cavities are enlarged and the walls increased in thickness (eccentric hypertrophy). The so-called concentric hypertrophy in which there is diminution of the size of the cavity with thickening of the walls is, as a rule, a post mortem change. The enlargement may affect the entire organ, or one side, or only one chamber. Naturally, as the left ventricle does the chief work the change is most frequently found here. Though its production is assisted by adequate nutrition, hypertrophy may appear even under conditions of starvation, given otherwise healthy organs. In the debilitated the limits to which hypertrophy may progress are small.

**HYPERTROPHY OF THE LEFT VENTRICLE ALONE**, or with general enlargement of the heart, is brought about by—

*Conditions affecting the heart itself:* (a) Disease of the aortic valve; (b) mitral insufficiency; (c) pericardial adhesions; (d) sclerotic myocarditis; (e) disturbed innervation with overaction, as in exophthalmic goitre, and as a result of the action of alcohol, in the “beer heart.” In all of these the work of the heart is increased. With the valve lesions the increase is due to increased intraventricular pressure with more blood to handle; in the case of the adherent pericardium and myocarditis, to direct interference with the proper contraction of the chambers.

*Conditions acting upon the blood-vessels:* (a) General arterio-sclerosis with hypertension, with or without renal disease, especially sclerosis of the aorta, the renal arteries, and the vessels of the splanchnic area; (b) all states of increased arterial tension induced by the contraction of the smaller arteries under the influence of spasm or toxic substances, which, as Bright suggested, “by affecting the minute capillary circulation, render great action necessary to send the blood through the distant subdivisions of the vascular system”; (c) prolonged muscular exertion, which increases the blood pressure in the arteries; (d) narrowing of the aorta, as in congenital stenosis.

**RIGHT VENTRICLE HYPERTROPHY** occurs with the following conditions:

(a) *Lesions of the mitral valve*, either incompetence or stenosis, which increase the resistance in the pulmonary vessels. (b) *Pulmonary lesions* with atheroma or obliteration of any number of vessels within the lungs, as in emphysema or fibrosis. (c) *Valvular lesions* on the right side occasionally cause hypertrophy in the adult, not infrequently in the fetus. (d) *Chronic valvular disease of the left heart and pericardial adhesions* are sooner or later associated with hypertrophy of the right ventricle.

In the auricles simple hypertrophy is never seen; there is always dilatation with hypertrophy. In the left auricle this develops in lesions of the mitral orifice, particularly stenosis. The right auricle hypertrophies when there is increased pressure in the lesser circulation, whether due to mitral stenosis or pulmonary lesions. Narrowing of the tricuspid orifice is a rare cause.

**Symptoms.**—There may be no complaint due to the hypertrophy and if associated with renal disease or arterio-sclerosis there may be a marked sense of well-being. If the cardiac defect is not fully compensated, the patient may complain of giddiness, headache, palpitation and some dyspnoea on exertion.

In hypertrophy of the *right auricle* the venous pulsation in the neck may be more evident, and an increase in dulness to the right of the sternum in

## DISEASES OF THE CIRCULATORY SYSTEM

the third and fourth interspaces may be detected. Hypertrophy of the *right ventricle* causes a slight bulging of the costal angle with a positive instead of a negative pulsation at this spot. The apex beat may be diffuse, as the enlarged right ventricle prevents the left ventricle from coming into contact with the chest wall. The venous pulsation in the neck is usually marked, and the first sound over the tricuspid area louder than normal. Hypertrophy of the *left auricle*, which is never unassociated with dilatation, may be detected occasionally by dullness toward the base of the left lung behind; it is easily diagnosed by the extension backward of the cardiac shadow in oblique illumination of the chest by the X-rays. Hypertrophy of the *left ventricle* is usually easy to diagnose. There is a forcible impulse at the apex beat, both visible and palpable, which may cause a movement of a large area of the chest wall. The apex beat, if there be only slight dilatation, is usually displaced downward, and is in the 6th and 7th spaces; but if the dilatation be marked, the apex beat becomes more diffuse and is well outside the nipple line in the 4th, 5th and 6th spaces. The first sound is usually marked and sometimes has a distinct booming quality. The second sound at the base is accentuated. The pulse is full and of high tension at the height of the ventricular impulse. The blood pressure is usually raised.

### II. DILATATION

As with other hollow muscular organs, the size of the chambers of the heart varies greatly within normal limits. Dilatation may be acute and quite transitory, as after severe muscular effort, or it may be chronic, in which case it is associated with hypertrophy. Not always, however; there is an extraordinary heart in the McGill College Museum showing a parchment like thinning of the walls with uniform dilatation of all the chambers; in places in the right auricle and ventricle only the epicardium remains. Dilatation is pathological only when excessive or permanent. Increase in capacity means increased work and in consequence hypertrophy to meet the demand.

**Etiology.**—Two important causes combine to produce dilatation—increased pressure within the cavities and impaired resistance, due to weakening of the muscular wall—which may act singly, but are often combined. A weakened wall due either to structural change in the muscle or to a diminution of its natural tonus may yield to a normal distending force.

(a) **HEIGHTENED ENDOCARDIAC PRESSURE** results from an increased amount of blood to be moved or an obstacle to be overcome. It does not necessarily cause dilatation; simple hypertrophy may follow, as in the early period of aortic stenosis, and the left ventricle hypertrophy in nephritis.

The size of the cardiac chambers varies in health. With slow action of the heart the dilatation is complete and fuller than it is with rapid action. Moderate exertion in a normal heart, or even prolonged exertion in a well-trained heart, lessens the heart size, but in conditions of ill health dilatation occurs. Physiologically, the limits of dilatation are reached when the chamber does not empty during systole. This occurs as an acute transient condition in severe exertion in an untrained or feeble condition.

There may be great dilatation of the *right heart*, as shown by the increased epigastric pulsation and increase in the cardiac dullness. The *safety valve*

## AFFECTIONS OF THE MYOCARDIUM

action of the tricuspid valves may come into play, relieving the lungs by permitting regurgitation into the auricle. With rest the condition is removed, but, if it has been extreme, the heart may suffer a strain from which it may recover slowly, or, indeed, the individual may never be able again to undertake severe exertion. In the process of training the getting wind, as it is called, is largely a gradual increase in the capability of the heart, particularly of the right chambers. A degree of exertion can be safely maintained in full training which is quite impossible under other circumstances, because, by a gradual process of what we may call physical education, the heart has strengthened its reserve force—widened enormously its limit of physiological work.

Endurance in prolonged contests is measured by the capabilities of the heart, which by increasing its tonus has increased its resistance to dilatation. We have no positive knowledge of the nature of the changes in the heart in this process, but it must be in the direction of increased muscular and nervous energy. The large heart of athletes may be due to the prolonged use of their muscles, but no man becomes a great runner or oarsman who has not naturally a capable if not a large heart. Master McGrath, the celebrated greyhound, and Eclipse, the race horse, both famous for endurance rather than speed, had very large hearts.

Excessive dilatation during severe muscular effort results in *heart-strain*. A man, perhaps in poor condition, calls upon his heart for extra work during the ascent of a mountain, and is seized with pain about the heart and a sense of distress in the epigastrium. He breathes rapidly for some time, is "puffed," as we say, but the symptoms pass off after a night's quiet. An attempt to repeat the exercise is followed by another attack, or dyspnoea may come on while at rest. For months such a man may be unfitted for severe exertion or he may be permanently incapacitated. In some way he has overstrained his heart and become "broken-winded." In such cases there was probably previous myocardial change. The "heart-shock" of Latham includes cases of this nature—sudden cardiac break-down during exertion, not due to rupture of a valve.

Acute dilatative heart weakness is seen in many conditions, as in Graves' disease, in paroxysmal tachycardia, in old myocardial cases following exertion, and in angina pectoris. There is usually a striking contrast between the wide cardiac impulse and the small, feeble, irregular pulse.

Dilatation occurs in all forms of *valve lesions*. In *aortic insufficiency* blood enters the left ventricle during diastole from the unguarded aorta and from the left auricle, and the quantity of blood at the termination of diastole subjects the walls to an extreme degree of pressure, under which they inevitably yield. In time they augment in thickness, and present the typical eccentric hypertrophy of this condition.

In *mitral insufficiency* blood which should have been driven into the aorta is forced into and dilates the auricle from which it came, and then in the diastole of the ventricle a large amount is returned from the auricle, and with increased force. In *mitral stenosis* the left auricle is the seat of greatly increased tension during diastole, and dilates as well as hypertrophies; the distention may be enormous. Dilatation of the *right ventricle* is produced by a number of conditions, which were considered under hypertrophy. All circum-

## DISEASES OF THE CIRCULATORY SYSTEM

stances, such as mitral stenosis, emphysema, etc., which permanently increase the tension in the pulmonary vessels cause its dilatation.

The dilatation and hypertrophy of beer drinkers also comes in this group, as it is brought about gradually by increased endocardial pressure.

(b) IMPAIRED NUTRITION OF THE HEART WALLS may lead to a diminution of the resisting power so that dilatation readily occurs.

The loss of tone due to parenchymatous degeneration or myocarditis in fevers may lead to a fatal condition of acute dilatation. It may occur in rheumatic, scarlet, typhus or typhoid fever, etc. The changes in the heart muscle which accompany acute endocarditis or pericarditis may lead to dilatation, especially in the latter disease. In anæmia and leukaemia the dilatation may be considerable. In fibrosis of the walls the yielding is always where this process is most advanced, as at the left apex. Under any of these circumstances the walls may yield with normal blood pressure.

Pericardial adhesions are a cause of dilatation, and we generally find in cases with extensive and firm union considerable hypertrophy and dilatation. There is usually here some impairment of the superficial layers of muscle. Dilatation is a common sequence of hypertrophy, usually after a long period. In many cases this occurs with the appearance of auricular fibrillation.

### III. MYOCARDIAL INSUFFICIENCY

**Etiology.**—With lessening of the muscular power of the heart the rapidity with which the blood circulates is diminished, and the tissues fail to receive their proper supply of oxygen and food, and to be adequately relieved of their waste products—this is cardiac failure. The same effect may be produced in another way. The amount of blood in the body is much less than the total capacity of the vascular bed, and an adequate blood supply is only kept up by a general constriction of arterioles which dam the blood in the arterial system, but if by any chance there is a general vaso-dilatation of the arterioles, especially those in the splanchnic area, the heart does not receive an amount of blood sufficient to supply the bodily needs, with the same effect on the organs as in certain forms of cardiac failure. This condition does not concern us here, but it must be mentioned to avoid the impression that all failure of the circulation means failure of the heart.

The failure in muscular power may affect any cavity singly or the whole heart. Weakness of the left ventricle fails to give proper filling of the arterial system and general anæmia of the tissues results. Failure of the left auricle means stasis in the lung vessels with deficient aëration of the blood, and a tendency to cedema of the lung or to effusion into the pleural cavity. Failure of the right heart gives cyanosis of the organs, dyspnoea at rest and on slight exertion, with stasis in the abdominal organs and cedema.

The reserve power with which the cardiac muscle is endowed disappears in heart failure. This reserve, greatest in youth, is increased by adequate nutrition, certain congenital endowments, and, apart from other defects, by hypertrophy. It is lessened by defects in the cardiac structure, gross or minute, by defective nutrition, by bacterial and other poisons, and with advancing years. We have at present no means of gauging this reserve power of the organ.

The failure may be sudden or slow, according to the kind and rapidity

## AFFECTIONS OF THE MYOCARDIUM

of the lesion which causes it. When the left ventricle fails the effect may vary from immediate death, through all forms of fainting, giddiness, sense of dissolution, to a mild sense of bodily or mental fatigue; when the right ventricle fails the effect varies from a sudden dyspnoea to a dyspnoea which comes on with slight exertion.

As to the actual condition in myocardial failure generally, it is by no means easy in all cases to say what has been the cause. The lesions to which the cardiac musculature is liable are described further on, yet there is a proportion of cases in which neither by post-mortem examination nor microscopic search can the cause of the failure be suggested. Disturbances in conduction, auricular fibrillation, or other changes in the rhythm are often responsible. One or more of the functions of the heart muscle may be affected.

The *blood pressure* in myocardial insufficiency shows no uniform figures. The systolic pressure may be high even in a failing heart. In serious degrees of myocardial affection it is usually low. In cases in which there has been a raised blood pressure, the maximum may be lower or higher than the normal for the patient. Probably in early stages of failure the heart is stimulated to put forth increased energy and the maximum pressure at the height of the beat slightly over-compensates the circulatory defect.

**ACUTE MYOCARDIAL INSUFFICIENCY.**—Causes: (a) Wounds of the heart, (b) spontaneous rupture or rupture of valves, (c) rapid pericardial effusion, (d) access of air to the chambers of the heart, as from operations on the neck or after exposure to a high atmospheric pressure, (e) large thrombi quickly formed in a heart cavity, (f) sudden interference with the coronary circulation, especially the left coronary artery, (g) mechanical interference from pressure on the trachea or larynx, as in strangulation, (h) acute infections, such as diphtheria or pericarditis, (i) certain poisons, such as cocaine, phosphorus, etc., (j) stimulation of the vagus nerve, its centre in the medulla, or its termination in the heart.

**CHRONIC MYOCARDIAL INSUFFICIENCY.**—Causes: (a) Lesions of the heart muscle, which will be described in more detail. *All cardiac failure is muscular.* The myocardium may be insufficiently nourished, as in the starvation atrophy of new growths or there may be recognizable lesions. One or more of the functions of the cardiac muscle may be interfered with without producing any changes that can be detected by the microscope. (b) Lesions of the valves. (c) Lesions affecting the vascular fields of the efferent arteries. Emphysema, chronic bronchitis, asthma, fibrosis of the lungs, chest deformities, and mitral disease produce an embarrassment of the right heart; atheroma of the aorta and arterio-sclerosis, especially of the splanchnic and renal area, produce failure of the left heart. (d) Over-exertion. (e) Certain poisons, such as alcohol (especially beer) and phosphorus. (f) Other causes, such as adherent pericardium and exophthalmic goitre.

**Anatomical Basis of Myocardial Insufficiency.**—I. LESIONS OF THE CORONARY ARTERIES.—A knowledge of the changes produced in the myocardium by disease of the coronary vessels gives a key to the understanding of many problems in cardiac pathology. The terminal branches of the coronary vessels are end arteries; that is, the communication between neighboring branches is through capillaries only. F. H. Pratt has shown that the vessels of Thebesius, which open from the ventricles and auricles into a system

## DISEASES OF THE CIRCULATORY SYSTEM

of fine branches and thus communicate with the cardiac capillaries and coronary veins, may be capable of feeding the myocardium sufficiently to keep it alive even when the coronary arteries are occluded. The blocking of one of these vessels leads usually to a condition known as—

(a) *Anæmic necrosis*, or white infarct. When this does not occur the reason may exist in (1) the existence of abnormal anastomoses, which take the coronary system out of the group of end arteries; or (2) the vicarious flow through the vessels of Thebesius and the coronary veins. The condition is most common in the left ventricle and in the septum, in the territory of distribution of the anterior coronary artery. The affected area has a yellowish white color, sometimes a turbid, parboiled aspect, at other times a grayish red tint. It may be somewhat wedge-shaped, more often it is irregular and projects above the surface. Microscopically the changes are characteristic. The nuclei disappear from the muscle fibres or undergo fragmentation. Leucocytes wander in from the surrounding tissue and may suffer disintegration. At a later stage a new growth of fibrous tissue is found in the periphery of the infarct which ultimately may entirely replace the dead fibres. In some instances there is complete transformation, and a firm white patch of hyaline degeneration may appear in the centre of the area. Rupture of the heart may be associated with anæmic necrosis.

(b) The second important effect of coronary artery disease is seen in the production of *fibrous myocarditis*. This may result from the gradual transformation of areas of anæmic necrosis. More commonly it is caused by the narrowing of a coronary branch in a process of obliterative endarteritis. Where the process is gradual, evidences of granulation tissue are often wanting, and any distinction between the necrotic muscle fibres and the new scar tissue is difficult to establish. J. B. MacCallum showed that the muscle fibres undergo a change the reverse of that of their normal development and lose their fibril bundles preliminary to their complete replacement by connective tissue. The sclerosis is most frequent at the apex of the left ventricle and in the septum, but may occur in any portion. In the septum and walls there are often streaks and patches seen only in carefully made serial sections. Hypertrophy of the heart is commonly associated with this degeneration. It is the invariable precursor of aneurism of the heart.

(c) *Sudden Death in Coronary Artery Disease*.—Complete obliteration of one coronary artery, if produced suddenly, is usually fatal. When induced slowly, by arterio-sclerosis at the orifice at the root of the aorta or by an obliterating endarteritis, the circulation may be carried on through the other vessel. Sudden death is not uncommon, owing to thrombosis of a vessel narrowed by sclerosis. *In medico-legal cases it is a point of primary importance to remember that this is one of the common causes of sudden death.* It should be carefully sought for, as it may be the sole lesion, except a general, sometimes slight, arterio-sclerosis. In the most extreme grade one coronary artery may be entirely blocked, with the production of extensive fibroid disease, and a main branch of the other also may be occluded.

(d) *Septic Infarcts*.—In pyæmia the smaller branches of the coronary arteries may be blocked with emboli which give rise to infectious or septic infarcts in the myocardium in the form of abscesses, varying in size from a pea to a pin's head. These may not cause any disturbance, but when large

## AFFECTIONS OF THE MYOCARDIUM

they may perforate into the ventricle or into the pericardium, forming what has been called acute ulcer of the heart.

II. ACUTE INTERSTITIAL MYOCARDITIS.—In some infectious diseases and in acute pericarditis the intermuscular connective tissue may be swollen and infiltrated with small round cells and leucocytes, the blood-vessels dilated, and the muscle fibres the seat of granular, fatty, and hyaline degeneration. Occasionally, in pyæmia the infiltration with pus cells is diffuse and confined chiefly to the interstitial tissue. Councilman described this condition of the heart wall in gonorrhœa, and demonstrated the gonococcus in the diseased areas. The commonest examples are found in diphtheria, typhoid fever, and acute endocarditis. The foci may be the starting points of patches of fibrous myocarditis.

III. FRAGMENTATION AND SEGMENTATION.—This condition was described by Renaut and Landouzy in 1877. Two forms are met with: (1) Segmentation. The muscle fibres have separated at the cement line. (2) Fragmentation. The fracture has been across the fibre itself, and perhaps at the level of the nucleus. Longitudinal division is unusual. Although the condition doubtless arises in some instances during the death agony, as in sudden death by violence, in others it would seem to have clinical and pathological significance. It is found associated with other lesions, fibrous myocarditis, infarction, and fatty degeneration. J. B. MacCallum distinguished a simple from a degenerative fragmentation. The first takes place in the normal fibre, which, however, shows irregular extensions and contractions. The second succeeds degeneration in the fibre. Hearts the seat of marked fragmentation are lax, easily torn, the muscle fibres widely separated, and often pale and cloudy.

IV. PARENCHYMATOUS DEGENERATION.—This is usually met with in fevers, or with endocarditis or pericarditis, and in infections and intoxications generally. It is characterized by a pale, turbid state of the cardiac muscle, which is general, not localized. Turbidity and softness are the special features. It is the softened heart of Laennec and Louis. Stokes speaks of an instance in which "so great was the softening of the organ that when the heart was grasped by the great vessels and held with the apex pointing upward, it fell down over the hand, covering it like a cap of a large mushroom." Histologically, there is a degeneration of the muscle fibres, which are infiltrated to a various extent with granules which resist the action of ether, but are dissolved in acetic acid. Sometimes the change in the fibres is extreme, and no trace of the striæ can be detected.

V. FATTY HEART.—Under this term are embraced fatty degeneration and fatty overgrowth or infiltration.

(a) *Fatty Degeneration*.—The frequency and importance of this condition have probably been greatly overestimated and the designation has been loosely used. Microscopically visible fat does not necessarily mean fatty degeneration, and it is a question whether fatty and fibrous changes can be differentiated. There is no question of its occurrence in phosphorus poisoning and pernicious anæmia, but in general a clinical diagnosis of fatty degeneration is most uncertain and there are no symptoms or signs peculiar to it.

(b) *Fatty Overgrowth or Infiltration*.—This is usually a simple excess of the normal subpericardial fat, to which the term *cor adiposum* was given by the older writers. In pronounced instances the fat infiltrates between the

## DISEASES OF THE CIRCULATORY SYSTEM

muscular substance and, separating the strands, may reach even to the endocardium. In corpulent persons there is always much pericardial fat. It forms part of the general obesity, and occasionally leads to dangerous impairment of the contractile power of the heart. Of 122 cases analyzed by Forchheimer there were 88 males and 34 females. Over 80 per cent. occurred between the fortieth and seventieth years. The entire heart may be enveloped in a thick sheeting of fat through which not a trace of muscle substance can be seen. On section the fat infiltrates the muscles, separating the fibres, and in some places there may be complete substitution of fat for the muscle substance. In rare instances the fat may be in the papillary muscles. The heart is usually much relaxed and the chambers are dilated.

VI. OTHER DEGENERATIONS.—(a) *Brown Atrophy*.—This is a common change in the heart muscle, particularly in chronic valvular lesions and in the senile heart. When advanced the color of the muscles is a dark red brown, and the consistence is usually increased. The fibres present an accumulation of yellow brown pigment chiefly about the nuclei. (b) *Amlyoid degeneration* occasionally occurs in the intermuscular connective tissue and in the blood vessels, not in the fibres. (c) The *hyaline transformation of Zenker* occurs in prolonged fevers. The affected fibres are swollen, homogeneous, translucent, and the striae are faint. (d) *Calcareous degeneration* occasionally occurs in the myocardium, and the muscle fibres are infiltrated with lime salts.

VII. FUNCTIONAL BASIS OF MYOCARDIAL INSUFFICIENCY.—The cause of insufficiency may be in disturbance of one or more of the functions of the heart muscle. In many cases it is probable that more than one is affected. In some cases the disturbance is temporary and complete recovery follows, as seen in auricular fibrillation or heart-block occurring with acute infectious diseases. Here inflammatory changes or disturbance from toxins may be responsible. The most striking common disturbance of function is seen in auricular fibrillation. The importance of structural changes which are usually responsible for the disturbances of function must be remembered.

**Symptoms.**—The symptoms of left-sided cardiac failure differ from those of the right side, and in each we may distinguish a number of forms, which, however, merge gradually the one into the other. Failure of the left ventricle is seen in its severest forms in the abrupt death stroke of angina pectoris, in the sudden syncope with sweating and heart pain of fibroid hearts, or in the attacks of Stokes-Adams disease. Less severe failure may be seen in athletes after a hard race, when vomiting and a feeling of dissolution are present—a form sometimes seen in angina, when it is liable to be mistaken for a gastrointestinal upset. The milder degrees show themselves in an inability to take much exercise or to do much mental work without a sense of great fatigue. Sudden and slow forms are also seen in failure of the right side. Subjected to a slight strain, great distress may come on, and one form of cardiac dyspnoea which attacks the patient at night is of this nature. The severer forms show an increasing inability to undergo slight extra exertion, such as mounting stairs, or hyperpnoea even when at rest in bed, in both of which there is usually some oedema of the feet, especially at night, if the patient is on his feet most of the day.

The symptoms complained of by patients with myocardial failure are as follows: (a) *Cardio-vascular system*: Pain in the cardiac area or extending

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to the shoulders and down the arms, a sense of weight in the præcordium; palpitation is a rare complaint. (b) Respiratory system: Dyspnoea at rest or on exertion, or orthopnoea, Cheyne-Stokes respiration, cough, loss of voice from pressure of a dilated left auricle on the left recurrent laryngeal nerve, hæmoptysis (from lung infarcts). (c) Central nervous system: sleeplessness, mental symptoms, delusions, melancholia, and especially toward the end stupor and drowsiness. (d) Cyanosis, pallor, œdema, and occasionally purpura in the lower limbs. (e) Alimentary system: The stasis in the abdominal organs in right heart failure produces loss of appetite, indigestion, flatulence, vomiting, constipation, diarrhoea, abdominal pain, hæmorrhoids, etc. (f) Renal system: The urine is scanty, high colored, and contains albumin.

Examination of the heart may reveal an apex-beat which is feeble, outside the nipple line, diffuse, and whose maximum intensity is not easily localized. The pulsation may be marked on inspection and cover a very wide area; arterial pulsation in the neck in left heart failure may be great; in right heart failure the jugular veins may be very dilated. On percussion the cardiac area may be much increased to the right or to the left, or both. On auscultation the sounds may be difficult to hear, or feebler than normal. With the muscular insufficiency there is often a relative insufficiency of the mitral and tricuspid valves, rarely of the aortic and pulmonic valves. If the patient is seen for the first time in an attack of decompensation, it may be impossible to decide whether a mitral insufficiency is relative (that is from inability of the valves to close the relaxed orifice) or due to organic valvular disease. In many elderly patients with arterio-sclerosis and hypertrophy, the murmur of relative insufficiency may be loud and intense. An exact diagnosis of the valvular lesion is of no great practical importance; the muscular insufficiency is the important factor. Gallop rhythm may be present. The pulse may show great variations; marked failure may exist with a full bounding pulse: more usually it is feeble with diminished tension; it may be irregular, intermittent, slow, or rapid. The vital capacity is much reduced from the normal 3,500-4,000 to 1,500-2,000 c. c. (J. H. Pratt).

When *dilatation* occurs there are gallop rhythm, shortening of the long pause, and a systolic murmur at the apex. Shortness of breath on exertion is an early feature in many cases, and anginal attacks may occur. There is sometimes a tendency to syncope, and the patient may wake from sleep in the early morning with an attack of severe dyspnoea. These "spells" may be associated with nausea and may alternate with others in which there are anginal symptoms. These are the cases, too, in which for weeks there may be mental symptoms. The patient has delusions and may even become maniacal. Toward the close Cheyne-Stokes breathing may occur. It was described in the following terms by John Cheyne (*Dublin Hospital Reports*, vol. ii, p. 221, 1818): "For several days his breathing was irregular; it would entirely cease for a quarter of a minute, then it would become perceptible, though very low, then by degrees it became heaving and quick, and then it would gradually cease again: this revolution in the state of his breathing lasted about a minute, during which there were about thirty acts of respiration." It is seen more frequently in arterio-sclerosis and uræmic states than in myocardial degeneration.

Fatty infiltration is a condition certain to exist in very obese persons.

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It produces no symptoms until the muscular fibre is so weakened that dilatation occurs. These patients may for years present a feeble but regular pulse; the heart sounds are weak and muffled, and a murmur may be heard at the apex. Attacks of dyspnoea are not uncommon, and the patient may suffer from bronchitis. The examination is often difficult because of the great increase in the fat, and it may be impossible to define the area of dulness.

*Thrombosis of the coronary arteries* occurs usually in middle-aged or elderly people. Their vessels are sclerotic, the blood pressure may be high and they may have had angina pectoris. The onset is usually very acute with sharp severe pain, which may be referred to the lower sternum, felt over the heart or referred to the epigastrium, preceded or accompanied by marked dyspnoea. There may be nausea and vomiting, cough and cyanosis, sweating and collapse. In patients who already had cardiac symptoms the onset may be less severe. The *pain* is a prominent feature and is persistent, sometimes with an occasional decrease in severity. It may radiate to the arms or neck, or around the body. It is often referred to the upper abdomen and, with vomiting and signs of collapse, an acute abdominal condition may be suspected. Some of the deaths attributed to "acute indigestion" belong here. The *dyspnoea* is constant and severe, and may be out of proportion to the physical findings. Marked *weakness* is usual, with anxiety and the picture of shock. The action of the *heart* is weak, with distant feeble sounds and a weak pulse. The area of dulness is increased, and a friction rub may be heard. The blood pressure is usually low. Pulmonary stasis or oedema, passive congestion of the kidney and general oedema may follow. Death may occur at once or after a few hours; some patients recover slowly, but as a rule are not able to do anything requiring much exertion. Recovery is most likely to follow thrombosis of smaller branches.

Following an *operation* a variety of disturbing conditions occur which should be carefully studied. Sometimes what is regarded as "acute dilatation" occurs with very rapid heart-action. Some are paroxysmal auricular tachycardia, auricular fibrillation or auricular flutter. Pressure on the vagus, or on the eyes may correct paroxysmal auricular flutter.

*Cardioptosis*.—This is found in those persons with visceroptosis. The heart is narrow, lies vertically and is low in position. It is found in the sub-normal type with arterial hypoplasia, a tendency to under-nutrition, and vaso-motor instability. Dilatation occurs readily with any slight infection or disturbance and is easily overlooked owing to the small size of the heart with which a normal extent of dulness represents enlargement. They respond quickly to rest and digitalis.

*Functional Tests*.—There are many of these, the principle being to have the patient perform certain exercises, such as hopping on one leg, bending over, etc., and then studying the circulatory response. The exercise chosen should be suitable for the age and habits of the patient. The extent of response (pulse rate, blood pressure) and the length of time it persists are important points. But every patient is constantly doing "functional tests" in his daily life, which careful inquiry should elicit. There is a reduction in the pulmonary *vital capacity* in cardiac insufficiency as a result of engorgement of the pulmonary vessels, or the presence of pleural effusion, infarct,

etc. The estimation of the vital capacity (spirometry) is a simple procedure, helpful in determining the extent of cardiac insufficiency.

We may group the cases of myocardial failure as follows:

(1) Those in which sudden death occurs with or without previous indications of heart-trouble. Sclerosis of the coronary arteries exists—in some with recent thrombus and white infarcts; in others with extensive fibroid disease. Some cases of sudden death may be due to deficiency in the coronary blood supply due to sudden lowering of the diastolic pressure. Many patients never complain of cardiac distress, but, as in the case of Chalmers, the celebrated Scottish divine, enjoy unusual vigor of mind and body.

(2) Cases in which there are cardiac arrhythmia, shortness of breath on exertion, attacks of dyspnoea, sometimes anginal attacks, collapse symptoms with sweats and slow pulse, and occasionally marked mental symptoms.

(3) Cases with general arterio-sclerosis and hypertrophy and dilatation of the heart. They are robust men of middle age who have worked hard and lived carelessly. Dyspnoea, cough, and swelling of the feet are the early symptoms, and the patient comes under observation either with a gallop rhythm, embryocardia, or an irregular heart with an apex systolic murmur of mitral insufficiency. Recovery from the first or second attack is the rule. It is a common form of myocardial disease.

**Diagnosis.**—The symptoms are the best aid in early diagnosis, as they give the first indication of loss of myocardial efficiency. The response to exertion is important, always remembering the normal ability of the patient. Shortness of breath, distress and precordial discomfort or pain, with increased cardiac rate are significant. The distress at first is on exertion, later when at rest or for some time after exertion. With the condition well marked there is rarely much difficulty.

**Prognosis.**—Each case must be judged on its own merits, special notice being taken of the age, probable origin, and anatomical basis of the insufficiency. With disturbance of rhythm the nature of this should be determined as it has an important bearing on the outcome. Permanent inversion of the T wave in leads I and II is of grave outlook. Alteration of the Q R S complex in all the derivations is serious. The outlook in affections of the myocardium in advanced life is extremely grave. Patients recover, however, in a surprising way from the most serious attacks, particularly those of the third group. The outlook in obstruction of the coronary arteries is very grave. Even if an attack is recovered from, the duration of life will not be long.

**Treatment.**—Some patients never come under treatment; the first are the final symptoms. Other patients with marked failure, if treated on general lines, recover quickly. Much more difficult is the management of those patients in whom there is marked disturbance of function as heart-block, auricular fibrillation or alternation of the heart.

The following are the measures in the treatment of myocardial failure:

(a) **REST.**—This is essential whenever there are symptoms of myocardial failure; the patient should be in bed in the most comfortable position. With severe dyspnoea he may be more comfortable in a chair. In the patient with myocardial disease who is without symptoms, a certain amount of rest is advisable, as many hours as possible in bed and an hour or two of rest

in the day. The amount of exertion should be carefully supervised. Rest of mind and freedom from anxiety are also important, if more difficult to secure.

(b) **DIET.**—In acute conditions it is usually well to limit this in amount, especially the fluids. With marked passive congestion liquid diet may be advisable; otherwise small amounts of simple food may be given at short intervals. In any case with dilatation it is well to limit the total daily intake of fluids to 1,500 c. c. A “dry diet” for a few days is sometimes useful. It is usually wise to avoid large amounts of carbohydrates but these patients often do well with considerable amounts of sugar.

(c) **THE RELIEF OF THE EMBARRASSED CIRCULATION.**

(1) *By Venesection.*—In cases of dilatation, from whatever cause, in mitral or aortic lesions or distention of the right ventricle in emphysema, when signs of venous engorgement are marked and when there is orthopnea with cyanosis, the abstraction of 20 to 30 ounces of blood is indicated. This is the occasion in which timely venesection may save the patient's life. It is particularly helpful in the dilated heart of arterio-sclerosis.

(2) *By Depletion through the Bowels.*—This is particularly valuable when dropsy is present. The salines are to be preferred; before breakfast from half an ounce to an ounce of Epsom salts may be given in concentrated form. This usually produces liquid evacuations. The compound jalap powder in half dram (2 gm.) doses, or elaterin (gr. 1/10 0.006 gm.) may be employed for the same purpose. Even when the pulse is feeble cathartics are well borne, and they deplete the portal system rapidly and efficiently.

(3) *Remedies Which Stimulate the Heart.*—Of these by far the most important is *digitalis*, which was introduced into practice by Withering. The indication for its use is insufficiency of the heart muscle, most especially when auricular fibrillation is present; it should not be given when there is good compensation. Broken compensation, no matter what valvular lesion is present, is the signal for its use. It slows and increases the force of the contractions. High blood pressure is not a contraindication to its use. The beneficial effects are best seen in cases of mitral disease with auricular fibrillation. On theoretical grounds it has been urged that its use is not so advantageous in aortic insufficiency, since it prolongs diastole and leads to greater distention. This need not be considered, and *digitalis* is just as serviceable in this as in any other condition associated with dilatation. It may be given in the form of the leaves, the tincture or the infusion; it is a matter of indifference if the drug is good. The dosage varies according to the gravity of the condition. In severe cases one to two drams (4-8 c. c.) of the tincture is given at once followed by 20 minims three times a day. In less severe cases 20-30 minims (1.3-2 c. c.) are given three times a day until a definite effect is produced. The weight of the patient should be considered. Free purgation is an advantage before *digitalis* is begun. The very large dosage should not be given as a routine and toxic effects should be avoided. The leaf (gr. i corresponding to 15 minims of the tincture) or the infusion (5 i-iv, 4-16 c. c.) may be employed in place of the tincture. The use of supposed “active principles” is not advised. If there is auricular fibrillation it is well to continue the use of *digitalis* indefinitely in the best dosage for that patient. The opinion that *digitalis* is of use in auricular fibrillation only does not seem justified, and

it will be found beneficial in other conditions, sometimes when there are but few symptoms and perhaps fewer signs of myocardial insufficiency.

Ill effects rarely follow digitalis. Toxic effects are seen in the production of nausea and vomiting, apparently due to a reflex action conveyed to the vomiting centre from the heart by the sympathetic fibres and probably by the vagus (Hatcher). There may be two beats of the heart to one of the pulse (heart-block) or alternation of the heart-beat. These subside on the withdrawal of the digitalis, and are rarely serious. There are patients who require and take digitalis uninterruptedly for years. When compensation has been re-established the drug may be omitted. When there is dyspnoea on exertion and cardiac distress, from 5 to 10 minims (0.3 to 0.6 c. c.) three times a day may be advantageously given for prolonged periods. In severe conditions and if there is vomiting it may be necessary to give digitalis or strophanthus intramuscularly. Some of the special fluid preparations of digitalis suitable for hypodermic use should be employed in doses of m 15-30 (1-2 c. c.). There is some risk in giving these drugs intravenously and this method should be used only in a severe emergency. Digitalis may be given by rectum, usually best in one large dose.

Of other remedies strophanthus alone is of service, but as its effect is uncertain when given by mouth it should be administered by intramuscular injection. Doses of 10 to 15 minims (0.6 to 1 c. c.) of the tincture or strophanthin gr. 1/200 (0.00032 gm.) are given and repeated once or twice at intervals of twenty-four hours. The intramuscular is safer than the intravenous administration. Convallaria, adonis, vernalis and sparteine are recommended as substitutes for digitalis, but their inferiority is so manifest that their use is rarely indicated. Drugs, such as caffeine, diuretin and theocin, are useful in case of oedema for their diuretic effect.

When anæmia is a marked feature iron should be given in full doses. Arsenic is an excellent substitute, and one or other, or both, should be administered. Strychnia may be given alone or in combination with digitalis in 1 or 2 drop doses of the 1 per cent. solution, or hypodermically in doses of 1/40-1/20 gr. (0.0016 to 0.003 gm.).

**Treatment of Special Symptoms.**—(a) DROPSY.—The improved circulation under the influence of digitalis hastens the interstitial lymph flow and favors resorption of the fluid. Cathartics, by depleting the portal circulation, promote the absorption of fluid from the lymph spaces and lymph sacs. To these measures the use of caffeine, diuretin and theocin may be added. In some cases, however, it is not relieved, and the legs may be punctured by ordinary aspirating needles, with rubber tubing attached, which are inserted and left for hours; they often drain away large amounts. This is better than scarification. Canton flannel bandages may be applied to the oedematous legs. With marked hydrothorax or ascites tapping is advisable before digitalis is given.

(b) DYSPNOEA.—The patients are usually unable to lie down and should have a comfortable bed-rest—if possible, one with lateral projections, so that in sleeping the head can be supported as it falls over. The dyspnoea is associated with dilatation, chronic bronchitis, or hydrothorax. The chest should be carefully examined, as hydrothorax is a common cause of shortness of breath. There are cases of mitral regurgitation with recurring hydrothorax,

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usually on the right side, which requires relief over long periods by tapping. For the nocturnal dyspnoea, particularly with restlessness, morphia is invaluable and may be given without hesitation. The value of the calming influence of opium in all conditions of cardiac insufficiency is not sufficiently recognized. There are instances of cardiac dyspnoea unassociated with dropsy, particularly in mitral valve disease, in which nitroglycerine or sodium nitrite is of service, given in increasing doses. They are especially useful in cases in which the pressure is high.

(c) PALPITATION AND CARDIAC DISTRESS.—With great hypertrophy and the throbbing which is so distressing in some cases of aortic insufficiency, aconite is of service in doses of from 1 to 3 drops every two or three hours. An ice bag over the heart is also of service. For the pains, often so marked in aortic lesions, iodide of potassium in 10-grain (0.6 gm.) doses, three times a day, or nitroglycerine may be tried. Small blisters are sometimes advantageous. An important cause of palpitation and distress is distention of the stomach or colon, for which suitable measures must be used.

(d) GASTRIC SYMPTOMS.—The cases of cardiac insufficiency which do badly and fail to respond to digitalis are often those in which nausea and vomiting are prominent features. The liver is often greatly enlarged; there is more or less stasis in the hepatic vessels, and but little can be expected of drugs until the venous engorgement is relieved. If the vomiting persists, it is best to stop food and give ice, small quantities of milk and lime water, and effervescing drinks. The bowels should be freely moved and drugs given hypodermically, if possible.

(e) COUGH AND HÆMOPTYSIS.—The former is almost a necessary concomitant of myocardial insufficiency, owing to engorgement of the pulmonary vessels and more or less bronchitis. It is allayed by measures directed rather to the heart than to the lungs. Hæmoptysis is sometimes beneficial. An army surgeon, who was invalided during the American Civil War on account of hæmoptysis, supposed to be due to tuberculosis, had for many years, in association with mitral insufficiency and enlarged heart, many attacks of hæmoptysis. He was sure that his condition was invariably better after an attack. It is rarely fatal, except in some cases of acute dilatation, and seldom calls for special treatment.

(f) SLEEPLESSNESS.—One of the most distressing features, even in the stage of compensation, is disturbed sleep. Patients may wake suddenly with throbbing of the heart, often in an attack of nightmare. Subsequently, when the compensation has failed, it is also a worrying symptom. The sleep is broken, restless, and frequently disturbed by frightful dreams. Sometimes a dose of spirit of chloroform with spirit of camphor will give a quiet night. The compound spirit of ether, Hoffmann's anodyne, though very unpleasant, is frequently a great boon in the period when compensation has partially failed and the patients suffer from restless and sleepless nights. Paraldehyde, chloral hydrate and barbital are sometimes serviceable, but it is best, if these fail, to resort to morphia without hesitation.

(g) RENAL SYMPTOMS.—With broken compensation and lowering of the tension, the urinary secretion is diminished, and the amount may sink to 5 or 6 ounces in the day. Digitalis and strophanthus usually increase the flow. A brisk purge may be followed by augmented secretion. The combination of

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digitalis, squill, and calomel is sometimes effective when digitalis alone has failed. Diuretin in doses of 15 grains (1 gm.) or theocin (gr. v, 0.3 gm.) three times a day is sometimes useful.

The DIET in chronic cardiac diseases may be difficult to regulate. Widal and others have shown that retention of the chlorides is an important factor in oedema. A milk diet favors their elimination, and in the intervals between attacks a salt free diet as far as possible should be used. Starchy foods and articles likely to cause flatulency should be forbidden.

In certain cases the plan recommended by Oertel is advantageous especially in those forms of heart weakness due to intemperance in eating and drinking and defective bodily exercise. The Oertel plan consists of three parts: First, the reduction in the amount of liquid. Oertel allows daily about 36 ounces of liquid, which includes the amount taken with the solid food.\* Free perspiration is promoted by bathing (if advisable, the Turkish bath), or even by pilocarpine. The second point is the diet. *Morning*—Cup of coffee or tea, with a little milk, about 6 ounces altogether. Bread, 3 ounces. *Noon*—Three to 4 ounces of soup, 7 to 8 ounces of roast beef, veal, game, or poultry, salad or a light vegetable, a little fish; 1 ounce of bread or farinaceous pudding; 3 to 6 ounces of fruit for dessert. No liquids at this meal, as a rule, but in hot weather 6 ounces of fluid may be taken. *Afternoon*—Six ounces of coffee or tea, with as much water. As an indulgence an ounce of bread. *Evening*—One or 2 soft-boiled eggs, an ounce of bread, perhaps a small slice of cheese, salad, and fruit: 10 to 12 ounces of fluid.

The most important element is graduated exercise, not on the level, but up hills of various grades. The distance walked each day is gradually lengthened. In this way the heart is systematically exercised and strengthened.

The Schott treatment consists in a combination of baths with exercises. The water has a temperature of from 82°-95° F., and is charged with CO<sub>2</sub>. The good effects are claimed to come from cutaneous excitation, induced by the mineral and gaseous constituents of the bath, and stimulation of the sensory nerves. Artificial baths can be used with various strengths of sodium chloride and calcium chloride. The exercises, resistance gymnastics, consist in slow movements executed by the patient and resisted by the operator. The best cases for this treatment are those with myocardial weakness. In the stage of broken compensation with dropsy, etc., and in marked arterio-sclerosis, it is not so suitable. The "neurotic heart" is often much benefited.

## III. ENDOCARDITIS

Inflammation of the lining membrane of the heart is usually confined to the valves, so that the term is practically synonymous with valvular endocarditis. It occurs in several forms—*acute*, characterized by the presence of vegetations with loss of continuity or of substance in the valve tissues; *sub-acute*; and so-called *chronic* (better *sclerotic*) with a slow sclerotic change, resulting in thickening, puckering, and deformity. A common form is an acute process occurring on sclerotic valves, already damaged—the so-called recurring endocarditis. The term "chronic endocarditis" is established by custom, but the process is sclerotic rather than inflammatory.

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### I. ACUTE ENDOCARDITIS

This occurs in rare instances as a primary, independent affection; but in the great majority of cases it is a complication in various infective processes, so that in reality the disease does not constitute an etiological entity. For convenience of description we speak of a simple or benign, and a bacterial, malignant, ulcerative or infective endocarditis.

**Etiology.**—SIMPLE ENDOCARDITIS does not constitute a disease of itself, but is invariably found with some other affection. In 330 cases of rheumatic fever at the Johns Hopkins Hospital there were 110 cases of endocarditis. Bouillaud first emphasized the frequency of the association of simple endocarditis with rheumatic fever. Before him, however, the association had been noticed. Tonsillitis may be complicated with endocarditis. Of the specific diseases of childhood it is not uncommon in scarlet fever, while it is rare in measles, diphtheria and small-pox. In typhoid fever it occurred in 6 of 1,500 cases.

In pneumonia both simple and malignant endocarditis occur. In 100 autopsies in this disease at the Montreal General Hospital there were 5 instances of the former. Among 61 cases of endocarditis studied bacteriologically in Welch's laboratory, pneumococci were found in 21 (Marshall). Of 517 fatal cases of acute endocarditis, 115 were with pneumonia—22.3 per cent. (E. F. Wells). Acute endocarditis may occur in pulmonary tuberculosis and was found in 12 cases in 216 post mortems.

In chorea simple warty vegetations are found on the valves in a large majority of all fatal cases, in 62 of 73 collected cases. There is no disease in which, post mortem, acute endocarditis has been so frequently found. The endocarditis of *syphilis* possibly belongs here. Little is known of the early stages but it probably occurs both in acute and recurrent forms. It is usually observed in late stages. And, lastly, simple endocarditis is met with in diseases associated with loss of flesh and progressive debility, as cancer, gout, and nephritis.

MALIGNANT, BACTERIAL OR INFECTIVE ENDOCARDITIS is met with: (a) As a primary disease of the lining membrane of the heart or of its valves.

(b) As a secondary affection in pneumonia, in various specific fevers, in septic processes of all sorts, and most frequently of all as an infection on old sclerotic valves. In a majority of all cases it is a local process in an acute infection. Congenital lesions are very prone to the severer forms of endocarditis, particularly affections of the orifice of the pulmonary artery and the margins of an imperfect ventricular septum (C. Robinson).

The existence of a primary endocarditis has been doubted; but there are instances in which persons previously in good health, without any history of affections with which endocarditis is usually associated, have been attacked by a severe infection. In one case death occurred on the sixth day and no lesions were found other than those of malignant endocarditis.

The simple endocarditis of rheumatic fever or of chorea rarely progresses into the malignant form. Of all acute diseases complicated with severe endocarditis pneumonia probably heads the list. Gonorrhoea is a much more common cause than has been supposed. The affection may complicate erysipelas, septicæmia (from whatever cause), and puerperal fever.

## ENDOCARDITIS

**Morbid Anatomy.**—SIMPLE ENDOCARDITIS is characterized by the presence on the valves or on the lining membrane of the chambers of minute vegetations, ranging from 1 to 4 mm. in diameter, with an irregular and fissured surface, giving to them a warty or verrucose appearance. Often these little cauliflower-like excrescences are attached by very narrow pedicles. They are more common on the left side of the heart than the right, and occur on the mitral more often than on the aortic valves. The vegetations are upon the line of closure of the valves—i. e., on the auricular face of the auriculo-ventricular valves, a little distance from the margin, and on the ventricular side of the sigmoid valves. It is rare to see any swelling or macroscopic evidence of infiltration of the endocardium in the neighborhood of even the smallest granulations, or of redness, even when they occur upon valves already the seat of sclerotic changes, in which capillary vessels extend to the edges. With time the vegetations may increase greatly in size, but in simple endocarditis the size rarely exceeds that mentioned above. Hirschfelder has shown experimentally that they may form with great rapidity, even in a few hours.

The vegetations consist of elements derived from the blood, and are composed of blood plates, leucocytes, and fibrin in varying proportions. At a later stage they appear as small outgrowths of connective tissue. The transition of one form into the other can often be followed. The process consists of a proliferation of the endothelial cells and the cells of the subendothelial layer which invade the fresh vegetation, and ultimately entirely replace it. The blood cells and fibrin disintegrate and are gradually removed. Even when the vegetation has been entirely converted into connective tissue it is often found at autopsy to be capped with a thin layer of fibrin and leucocytes.

Micro-organisms are generally, even if not invariably, found associated with the vegetations. They tend to be entangled in the granular and fibrillated fibrin or in the older ones to cap the apices.

**SUBSEQUENT CHANGES.**—(a) The vegetations may become organized and the valve restored to a normal state (?). (b) The process may extend, and a simple may become an ulcerative endocarditis. (c) The vegetations may be broken off and carried in the circulation to distant parts. (d) The vegetations become organized and disappear, but they initiate a nutritive change in the valve tissue which ultimately leads to sclerosis, thickening, and deformity. The danger in simple endocarditis is not immediate, but remote, and consists in the process which results in sclerosis of the valves.

In *subacute bacterial* endocarditis the lesion, which may be large or small, rarely ulcerates and if this occurs it is usually in the aortic valves. It involves the mitral more often than the aortic orifice and very rarely the right heart. The chordæ tendineæ are often involved and there may be mural endocarditis. If healing occurs there is fibroid change, possibly with calcareous deposit.

**MALIGNANT OR BACTERIAL ENDOCARDITIS.**—Practically in every case of this form vegetations are present. In it the loss of substance in the valve is more pronounced, the deposition—thrombus formation—from the blood is more extensive, and the organisms are present in greater number and often show increased virulence. This form is often found in heart valves already the seat of chronic proliferative and sclerotic changes.

There is much loss of substance, which may be superficial and limited

to the endocardium, or, what is more common, it involves deeper structures, and not very infrequently leads to perforation of a valve, the septum, or even of the heart itself. The affected valve shows necrosis, with more or less loss of substance; the tissue is devoid of preserved nuclei and presents a coagulated appearance. Upon it a mixture of blood plates, fibrin and leucocytes enclosing masses of micro-organisms is found. The subjacent tissue often shows sclerotic thickening and always infiltration with exuded cells.

**PARTS AFFECTED.**—The following figures, taken from the Goulstonian lectures (Osler) give an approximate estimate of the frequency with which in 209 cases different parts of the heart were affected in malignant endocarditis: Aortic and mitral valves together, in 41; aortic valves alone, in 53; mitral valves alone, in 77; tricuspid in 19; the pulmonary valves in 15; and the heart walls in 33. In 9 instances the right heart alone was involved, in most cases the auriculo-ventricular valves.

*Mural* endocarditis is seen most often at the upper part of the septum of the left ventricle. Next in order is endocarditis of the left auricle on the postero-external wall. The vegetations may extend along the intima of the pulmonary artery into the lung. A common result of the ulceration is the production of valvular aneurism. In three-fourths of the cases the affected valves present old sclerotic changes. The process may extend to the aorta, producing extensive endarteritis with multiple acute aneurisms.

**ASSOCIATED LESIONS.**—The associated changes are those of the primary disease, those due to embolism, and the changes in the myocardium. In the endocarditis of septic processes there is the local lesion—an acute necrosis, a suppurative wound, or puerperal disease.

The changes due to *embolism* are striking, but it is remarkable that in some instances, even with endocarditis of a markedly ulcerative character, there may be no trace of embolic processes. The infarcts may be few in number—only one or two, perhaps, in the spleen or kidney—or they may exist in hundreds throughout various parts of the body. They may present the ordinary appearance of red or white infarcts of a suppurative character. They are most common in the spleen and kidneys, though they may be numerous in the brain, and in many cases are abundant in the intestines. In right sided endocarditis there may be infarcts in the lungs. In many of the cases there are innumerable miliary abscesses. Acute suppurative meningitis was met with in 5 of the 23 Montreal cases and in over 10 per cent. of 209 cases analyzed in the literature. Acute suppurative parotitis may occur. Lastly, the accompanying *myocarditis* plays an important rôle. The valvular insufficiency in acute endocarditis is probably not due to the row of little vegetations, but to the *myocarditis*, which interferes with the proper closure of the orifice.

**Indeterminate Forms.**—Under this heading Libman includes: (1) Atypical forms of verrucous endocarditis and (2) cases described as terminal endocarditis (diabetes, nephritis, etc.).

**Bacteriology.**—No distinction in the micro-organisms found in the two forms of endocarditis can be made. In both, cocci—streptococci, staphylococci, pneumococci, and gonococci—are the most frequent. More rarely, especially in simple vegetative endocarditis, the bacilli of tuberculosis, typhoid fever, and anthrax have been encountered. The colon bacillus has been found, and Howard described a case of malignant endocarditis due to an attenuated

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form, of the diphtheria bacillus. Marshall in 61 cases found the pneumococci in 21, streptococci alone or with other bacteria in 26, staphylococcus pyogenes aureus in 12. The meningococcus may cause endocarditis. Combined infections are not uncommon. In the subacute bacterial form the *Streptococcus viridans* is the common organism (Libman). As a rule no organisms are found in the simple endocarditis in chronic diseases, as tuberculosis, nephritis, etc. They may have been present and died out.

**Symptoms.**—The clinical course and the physical signs of SIMPLE ENDOCARDITIS are not in any respect characteristic. The great majority of the cases are latent and there is no indication of cardiac mischief. Endocarditis is frequently found post mortem in persons in whom it was not suspected during life. There are certain features, however, by which its presence is indicated with a degree of probability. The patient, as a rule, does not complain of any pain or cardiac distress. In rheumatic fever, for example, the symptoms to excite suspicion would be increased rapidity of the heart, perhaps slight irregularity, and an increase in the fever, without aggravation of the arthritis. Rows of tiny vegetations on the mitral or on the aortic segments seem a trifling matter to excite fever, and it is difficult in the endocarditis of febrile processes to say definitely in every instance that an increase in the fever depends upon this complication; but a study of the recurring endocarditis shows that the process may be associated, for weeks or months, with fever ranging from 100° to 103°. Palpitation may be a marked feature and is a symptom upon which certain authors lay great stress.

The *diagnosis* rests upon physical signs, which are notoriously uncertain. The presence of a murmur in a case of fever is often taken as proof of the existence of endocarditis—a common mistake due to the fact that a murmur is common to it and to a number of other conditions. At first there may be only a slight roughening of the first sound, which may gradually increase to a distinct murmur. The apex systolic murmur is often the result of *myocarditis*. It may not be present in the endocarditis of such chronic maladies as tuberculosis and carcinoma, since in them the muscle involvement is less common (Krehl). Reduplication and accentuation of the pulmonic second sound are frequently present. An aortic diastolic murmur is good evidence of endocarditis.

It is difficult to give a satisfactory clinical picture of MALIGNANT ENDOCARDITIS because the modes of onset are so varied and the symptoms so diverse. Arising in the course of some other disease, there may be simply an intensification of the fever or a change in its character. In a majority of the cases there are present certain general features, such as irregular pyrexia, sweating, delirium, and gradual failure of strength.

*Embolic* processes may give special features, such as delirium, coma, or paralysis from involvement of the brain or its membranes, pain in the side and local peritonitis from infarction of the spleen, bloody urine from implication of the kidneys, impaired vision from retinal hæmorrhage and suppuration, and even gangrene in various parts.

Various clinical forms have been described depending on the most marked features, such as septic, typhoid, and cerebral. The classification given by Libman into acute and sub-acute bacterial forms is more satisfactory. (1) *Acute Bacterial*. This is due usually to hæmolytic streptococci, pneumo-

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cocci, staphylococci, or gonococci and represents the form termed malignant endocarditis. (2) *Subacute Bacterial*. These are nearly always due to non-hæmolytic streptococci (*Streptococcus viridans*). The *B. influenza* is found in a few cases and the gonococcus rarely.

The "septic form" usually occurs in connection with an external wound, the puerperal process, acute necrosis or gonorrhœa. There are rigors, sweats, irregular fever, and all the signs of septic infection. The heart symptoms may be completely masked by the general condition, and attention called to them only by the occurrence of embolism. In many cases the features are those of a severe septicæmia, and the organisms may be isolated from the blood. Optic neuritis is not uncommon, and was present in 15 cases of chronic septic endocarditis examined by Faulkner, and in four of these recurrent retinal hæmorrhages were present.

The "typhoid form" is characterized by a less irregular fever, early prostration, delirium, somnolence, and coma, diarrhœa, sweating, which may be of a drenching character, petechial and other rashes, and occasionally parotitis.

The heart symptoms may be completely overlooked and in some instances careful examination has failed to discover a murmur.

In what may be termed the "cerebral form" the clinical picture may simulate a meningitis. There may be acute delirium or, as in three of the Montreal cases, the patient may be brought into the hospital unconscious.

Certain special features may be mentioned. The fever is not always of a remittent type, but may be high and continuous. Petechial rashes are common and the similarity is strong to certain cases of typhoid and cerebro-spinal fever. In one case the disease was thought to be hæmorrhagic small-pox. There may be small hæmorrhages in the palms and soles. Erythematous rashes are not uncommon. The sweating may be most profuse, exceeding that in pulmonary tuberculosis. Diarrhœa is not necessarily associated with embolic lesions in the intestines. Jaundice has been observed, and some cases have been mistaken for acute yellow atrophy. The heart symptoms may be entirely latent and are not found unless a careful search be made. Cases with chronic valve disease usually present no difficulty in diagnosis.

The course is varied, depending largely upon the nature of the primary trouble. Except in the disease grafted upon chronic valvulitis the course is rarely beyond five or six weeks. The most rapidly fatal case on record is described by Eberth, the duration of which was scarcely two days.

**SUBACUTE BACTERIAL ENDOCARDITIS.**—Due particularly to the work of Libman we recognize that these cases are much more common than was supposed. Organisms of the *Streptococcus viridans* group are found. A special feature is that the patients may become bacteria-free. The clinical picture is very varied: the prominent signs given by Libman are: (1) endocarditis; (2) multiple arterial embolism; (3) bacteria in the blood; and (4) fever. Certain other features are important: (a) marked progressive *anæmia*; (b) hæmorrhagic features, especially petechial; some cases of chronic purpura probably belong here; (c) painful *cutaneous nodes*, red raised spots on the skin of the hands and feet lasting a few days (Osler nodes); (d) marked renal disease, due to embolism (see embolic focal nephritis); (e) marked splenic enlargement; (f) *arthritis*; (g) symptoms from the nervous system,

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cerebral or meningeal; (h) *clubbing* of the fingers; and (i) brown *pigmentation* of the face. The renal changes are especially in the glomeruli and are embolic. Renal insufficiency is a common cause of death. The anæmia is of the secondary type and usually the leucocytes are normal or diminished. Tenderness over the sternum is a special feature and may be most marked in the bacteria-free stage.

As to the *course*, Libman's studies show that the patients with a positive blood culture nearly all die. If positive are followed by negative cultures, death usually occurs within a few months. In patients in whom the cultures are negative some are otherwise like those with positive cultures, others gradually lose the fever but die later, and in still others, fever may be absent for long periods. In the last group there has usually been previous endocarditis and anæmia and renal disease are common. The duration may be from a few months to two years. There are all grades of severity and some cases are very mild.

In many cases the process is engrafted on an old, sometimes an unrecognized, valve lesion. At first fever may be the only feature; in a few cases there are chills at the onset or recurring chills may arouse the suspicion of malaria. The patient may keep at work with a daily rise of temperature and perhaps an occasional sweat. The heart features may be overlooked. The murmur of the old valve lesion may show no change, and even with the most extensive disease of the mitral cusps the heart's action may be little disturbed. For months—six, eight, ten, even thirteen—fever and progressive weakness may be the most marked features. It is in such cases that the embolic phenomena are of special aid in diagnosis. Hematuria or tender skin nodes may be suggestive. The cutaneous nodes on the hands or feet occur as small painful red areas. With involvement of the aortic segments the signs of a progressive lesion are more common.

**Diagnosis.**—In many cases this is very difficult; in others, with marked embolic symptoms, it is easy. From simple endocarditis it is readily distinguished, though confusion occasionally occurs in the transitional stage, when a simple is developing into a malignant form. The constitutional symptoms are graver, the fever is higher, rigors are common, and septic symptoms occur. Perhaps a majority of the cases not associated with puerperal processes or bone disease are confounded with typhoid fever. A differential diagnosis may be impossible until blood cultures or specific reactions are positive. Points which may guide us are: The more abrupt onset in endocarditis, the absence of any regularity of the pyrexia in the early stage and the cardiac pain. Oppression and shortness of breath may be early symptoms. Rigors, too, are not uncommon. There is a marked leucocytosis in infective endocarditis. Between pyæmia and malignant endocarditis there are practically no differential features, for the disease really constitutes an *arterial pyæmia* (Wilks). In acute cases resembling the fevers the diagnosis of typhus, typhoid or cerebro-spinal fever may be made. The intermittent pyrexia, for weeks or months, has led to the diagnosis of malaria, but this disease can be excluded by the blood examination. Blood cultures aid greatly and are necessary for an etiological diagnosis. In the *subacute* form, a period of study may be necessary, especially if blood cultures are negative, and particular watch should be kept for the special features of this form.

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The cases usually terminate fatally. The instances of recovery are the subacute forms and the recurring endocarditis on old sclerotic valves.

**Treatment.**—We know no measures by which in rheumatic fever, chorea, or the eruptive fevers endocarditis can be prevented. As many cases arise, particularly in children, in mild forms of these diseases, it is well to insist upon rest and quiet, and to bear in mind that of all complications an acute endocarditis, though in its immediate effects mild, is ultimately serious. This is enforced by the observations of Sibson that on a system of absolute rest the proportion of cases of rheumatic fever attacked by endocarditis was less than of those who were not so treated. It is doubtful in rheumatic fever whether the salicylates have an influence in reducing the liability to endocarditis. Considering the extremely grave after-results of simple endocarditis in children, the question arises whether it is possible to do anything to avert the onset of progressive sclerosis of the affected valve. Caton recommends a systematic plan of treatment: (1) Prolonged rest in bed for three months; (2) a series of small blisters over the heart; and (3) iodide of potassium in moderate doses for many months. If there is much vascular excitement aconite may be given and an ice bag placed over the heart. The treatment of malignant endocarditis is practically that of septicæmia—useless and hopeless in a majority of the cases. Blood cultures should be taken as soon as possible and a vaccine prepared. Horder and others have reported good results. Personally we have not seen a successful case. Capps advises the use of sodium cacodylate (gr. i-iv, 0.06-0.24 gm. daily, usually given intravenously) in the subacute bacterial form. The drug is given until there is a strong garlic odor to the breath.

### II. SCLEROTIC CHRONIC ENDOCARDITIS

**Definition.**—A sclerosis of the valves leading to shrinking, thickening, and adhesion of the cusps, often with the deposition of lime salts, with shortening and thickening of the chordæ tendinæ, leading to insufficiency and to narrowing of the orifice. It may be primary, but is more often secondary to acute endocarditis, particularly the rheumatic form.

**Etiology.**—It is a mistake to regard every case of sclerotic valve as a sequel to an acute endocarditis. Long ago Roy and Adami called attention to the possibility that sclerosis of the valve segments might be a sequel of high pressure. The preliminary endocarditis may be a factor in weakening the valve, the progressive thickening of which may be a direct consequence of the strain. As age advances the valves begin to lose their pliancy, show slight sclerotic changes and foci of atheroma and calcification. The toxins of the specific fevers may initiate the change. A very important factor in the case of the aortic valves is *syphilis*. The strain of prolonged and heavy muscular exertion may play a part. In the aortic segments it may be only the valvular part of a general arterio-sclerosis.

The frequency with which sclerotic endocarditis is found may be gathered from the following figures: In from 12,000 to 14,000 autopsies in Dresden, Würzburg, and Prague, the percentage ranged from four to nine. The relative frequency of involvement of the valves is thus given in the collected statistics of Parrot: The mitral orifice in 621, aortic in 380, tricuspid in

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46, and the pulmonary in 11. This gives 57 instances in the right to 1,001 in the left heart.

**Morbid Anatomy.**—Vegetations in the form in which they occur in acute endocarditis are not present. In the early stage, the edge of the valve is a little thickened and perhaps presents a few small nodular prominences, which in some cases may represent the healed vegetations of the acute process. In the aortic valves the tissue about the corpora Arantii is first affected, producing a slight thickening with an increase in the size of the nodules. The substance of the valve may lose its translucency, and the only change noticeable is a grayish opacity and a slight loss of its delicate tenuity. In the auriculo-ventricular valves these early changes are seen just within the margin and here it is not uncommon to find swellings of a grayish red, somewhat infiltrated appearance, almost identical with the similar structures on the intima of the aorta in arterio-sclerosis. Even early there may be seen yellow or opaque white subintimal fatty degenerated areas. As the sclerotic changes increase, the fibrous tissue contracts and produces thickening and deformity of the segment, the edges of which become round, curled, and incapable of that delicate apposition necessary for perfect closure. An aortic valve may be narrowed one fourth or even one third across its face, the most extreme grade of insufficiency being induced without any special deformity and without any narrowing of the orifice. In the auriculo-ventricular segments a simple process of thickening and curling of the edges of the valves, inducing a failure to close without forming any obstruction to the normal course of the blood-flow, is less common. Still, we meet with instances at the mitral orifice, in which the edges of the valves are curled and thickened, so that there is extreme insufficiency without any material narrowing of the orifice. More frequently, as the disease advances, the chordæ tendinæ become thickened, first at the valvular ends and then along their course. The edges of the valves at their angles are gradually drawn together and there is a narrowing of the orifice, leading in the aorta to more or less stenosis and in the mitral orifice to constriction. Finally, in the sclerotic and necrotic tissues lime salts are deposited and may even reach the deeper structures of the fibrous rings, so that the entire valve becomes a dense calcareous mass with scarcely a remnant of normal tissue. The chordæ tendinæ may gradually become shortened, greatly thickened, and in extreme cases the papillary muscles are implanted directly upon the sclerotic and deformed valve. The apices of the papillary muscles usually show marked fibroid change.

In all stages the vegetations of simple endocarditis may be present, and the severer, ulcerative forms often attack these sclerotic valves.

Chronic *mural* endocarditis produces cicatricial-like patches of a grayish white appearance which are sometimes seen on the muscular trabeculæ of the ventricle or in the auricles. It often occurs with myocarditis.

The endocarditis of the fetus is usually of the sclerotic form and involves the valves of the right more frequently than those of the left side.

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## IV. CHRONIC VALVULAR DISEASE

### GENERAL INTRODUCTION

**Effects of Valve Lesions.**—The general influence on the work of the heart may be briefly stated as follows: The sclerosis induces insufficiency or stenosis, which may exist separately or in combination. The narrowing retards in a measure the normal outflow and the insufficiency permits the blood current to take an abnormal course. The result in the former case is difficulty in the

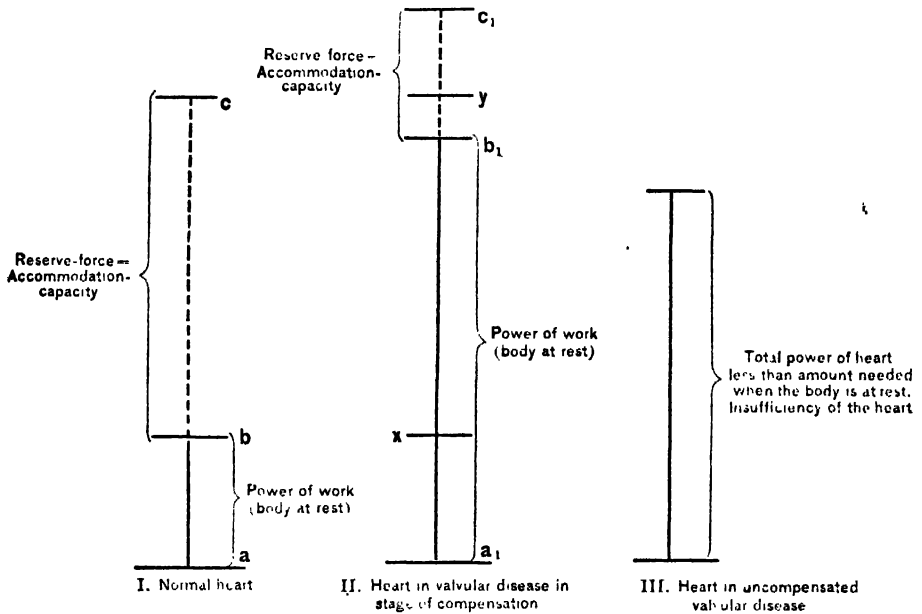


FIG. 12.—DIAGRAMMATIC PRESENTATION SHOWING THE FORCE OF THE HEART FOR WORK UNDER NORMAL CONDITIONS AND IN VALVULAR LESIONS.

expulsion of the contents of the chamber through the narrow orifice; in the other, the overfilling of a chamber by blood flowing into it from an improper source as in mitral insufficiency, when the left auricle receives blood both from the pulmonary veins and from the left ventricle. In both instances the effect is dilatation of a chamber, and to expel the normal amount of blood from a dilated chamber a relatively greater amount of energy is required, which by various adjustments the muscle is stimulated to do.

The cardiac mechanism is fully prepared to meet ordinary grades of dilatation which constantly occur during sudden exertion. A man at the end of a hundred yard race has his right chambers greatly dilated and his reserve cardiac power worked to its full capacity. The slow progress of sclerotic changes brings about a gradual, not an abrupt, insufficiency, and the moderate dilatation which follows is at first overcome by the exercise of the ordinary reserve strength of the heart muscle. Gradually a new factor is introduced. The constant increase in the energy put forth by the heart is a

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stimulus to the muscle fibres to increase in bulk and probably also in number; the heart hypertrophies, and the effect of the valve lesion becomes, as we say, *compensated*. The equilibrium of the circulation is in this way maintained.

The nature of the process is illustrated in the accompanying diagram, from Martius. The perpendicular lines in the figures represent the power of work of the heart. While the muscle in the healthy heart (Diagram I) has at its disposal the maximal force,  $a c$ , it carries on its work under ordinary circumstances (when the body is at rest) with the force  $a b$  and  $b c$  is the reserve force by which the heart accommodates itself to greater exertion.

If there be a gross valvular lesion, the force required to do the ordinary work of the heart (at rest) becomes very much increased (Diagram II). But in spite of this enormous call for force, insufficiency of the muscles does not necessarily result, for the working force required is still within the limits of the maximal power of the heart,  $a_1 b_1$  being less than  $a_1 c_1$ . The muscle accommodates itself to the new condition by making its reserve force mobile. If nothing further occurred, this could not be permanently maintained, for there would be left over for emergencies only the small reserve force  $b_1 y$ . Even at rest the heart would be using continuously almost its entire maximal force. Any slight exertion requiring more extra force than that represented by the small value  $b_1 y$  (say the effort required in walking or on going upstairs) would bring the heart to the limit of its working power, and dyspnoea would appear. Such a condition does not last long. The working power of the heart gradually increases. More and more exertion can be borne without causing dyspnoea, for *the heart hypertrophies*. Finally, a new, more or less permanent condition is attained, in that the hypertrophied heart possesses the maximal force,  $a_1 c_1$ . Owing to the increase in volume of the heart muscle, the total force of the heart is greater *absolutely* than that of the normal heart by the amount  $y c_1$ . It is, however, *relatively* less efficient, for its reserve force is much less than that of the healthy heart. Its capacity for accommodating itself to unusual calls upon it is permanently diminished.

Turning to the disturbances of compensation, it is to be distinctly borne in mind that any heart, normal or diseased, can become insufficient whenever a call upon it exceeds its maximal working capacity. The liability to such disturbance will depend, above all, upon the accommodation limits of the heart—the less the width of the latter, the easier will it be to go beyond the heart's efficiency. A comparison of Diagrams I and II will immediately make it clear that the heart in valvular disease will become insufficient much earlier than the heart of a healthy individual. The heart in valvular disease, on account of its small amount of reserve force, has to do maximal or nearly maximal work far more frequently than the normal heart. The power of the heart may become decreased to the amount necessary simply to carry on its work when the body is at rest, or it may cease to be sufficient even for this. The reserve force gained through the compensatory process may be entirely lost (Diagram III). If the loss be only temporary, the exhausted heart muscle quickly recovering, the condition is spoken of as a "disturbance of compensation." The term "loss of compensation" is reserved for the condition in which the disturbance is continuous.

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### AORTIC INSUFFICIENCY

This best-defined and most easily recognized of valvular lesions was first carefully studied by Corrigan, whose name it sometimes bears.

**Etiology and Morbid Anatomy.**—It is more frequent in males than in females, affecting chiefly men at the middle period of life. The ratio which it bears to other valve diseases has been given as from 30 to 50 per cent.

There are six groups of cases: I. Those due to *congenital malformation*, particularly fusion of two of the cusps—most commonly those behind which the coronary arteries are given off. It is possible that such aortic valves may be competent but a great danger is the liability of the malformed segments to sclerotic endocarditis. Of 17 cases all presented sclerotic changes, and the majority had, during life, the clinical features of chronic heart disease.

II. *The Endocarditic Group.*—Endocarditis may produce an acute insufficiency by ulceration and destruction of the valves; the aortic valves may be completely eroded away. The valvulitis of rheumatic fever, while more rarely aortic, is common enough, and the insufficiency is caused by nodular excrescences at the margins or in the valves, which may ultimately become calcified; more often it induces a slow sclerosis of the valves with adhesions, causing also some degree of narrowing.

III. *Syphilis.*—This is the most important cause, certainly in young adults and middle-aged patients. In a series of 200 cases in the Jefferson Hospital, Regester found about 60 per cent. due to syphilis. The spirochaetes may be found in the valves. The process frequently involves the aorta also. In some cases it causes a localized process at the root of the aorta which involves the valves secondarily or causes dilatation of the aortic ring with relative insufficiency. Some of the supposed cases of cure of aortic endocarditis may be instances of the latter.

IV. *The Arterio-sclerotic Group.*—A common cause of insufficiency is a slow, progressive sclerosis of the segments, resulting in a curling of the edges. It may be associated with general arterio-sclerosis. The condition of the valves is such as has been described in chronic endocarditis. It may be noted, however, how slight a grade of curling may produce serious insufficiency. Associated with the valve disease is, in a majority of cases, a more or less advanced arterio-sclerosis of the arch of the aorta, one serious effect of which may be a narrowing of the orifices of the coronary arteries. The sclerotic changes are often combined with atheroma which may exist at the attached margin of the valves without inducing insufficiency. In other instances insufficiency may result from a calcified spike projecting from the aortic attachment into the body of the valve, and so preventing its proper closure. Anatomically one can usually recognize the arterio-sclerotic variety by the smooth surface, the rounded edges, and the absence of excrescences.

V. Insufficiency may be induced by *rupture of a segment*—a very rare event in healthy valves, but not uncommon in disease, either from excessive effort during heavy lifting or from the ordinary strain on a valve eroded and weakened by ulcerative endocarditis.

VI. *Relative insufficiency*, due to dilatation of the aortic ring and adjacent arch, is very infrequent. It occurs in extensive sclerosis of the ascending

portion of the arch with great dilatation just above the valves. The valve segments are usually involved with the arterial coats, but the changes in them may be slight. In aneurism just above the aortic ring relative insufficiency of the valve may be present. Whether aortic insufficiency occurs from dilatation of the left ventricle has been much discussed—a relative incompetency similar to that which occurs at the pulmonary orifice. Cases are reported in which transient diastolic murmurs have occurred with dilatation of the heart, of which Anders reported and collected corroborative cases. J. B. MacCallum, whose untimely death was a great loss to science, described a sphincter-like band of muscle encircling the opening of the left ventricle into the aorta, and the relaxation of this ring muscle may be associated with insufficiency of the valve.

Insufficiency may be combined with various grades of narrowing, particularly in the endocarditic group. In a majority of the cases of the arterio-sclerotic form there is no stenosis but with aortic stenosis there is almost without exception some grade, however slight, of insufficiency.

**Effects.**—The direct effect of aortic insufficiency is the regurgitation of blood from the artery into the ventricle, causing an overdistention of the cavity and a reduction of the blood column; that is, a relative anæmia in the arterial tree. The amount returning varies with the size of the opening. The double blood flow into the left ventricle causes dilatation of the chamber, and finally hypertrophy, the grade depending upon the lesion. In this way the valve defect is compensated, and, as with each systole a larger amount of blood is propelled into the arterial system, the regurgitation of a certain amount during diastole does not, for a time at least, seriously impair the nutrition of the peripheral parts. For a time at least there is little or no resistance offered to the blood flow from the auricle—the ventricle accommodates itself readily to the extra amount, and there is no disturbance in the lesser circulation. In acute cases with rapid destruction of the segments, there may be the most intense dyspnoea and even profuse hæmoptysis.

In this lesion dilatation and hypertrophy reach their most extreme limit. The heaviest hearts on record are described in this affection. The so-called bovine heart, *cor bovinum*, may weigh 35 or 40 ounces, or, as in a case of Dulles's, 48 ounces. The dilatation is usually extreme and is in marked contrast to the condition in pure aortic stenosis. The papillary muscles may be greatly flattened. The mitral valves are usually not seriously affected, though the edges may present slight sclerosis, and there is often relative insufficiency, owing to distention of the mitral ring. Dilatation and hypertrophy of the left auricle are common, and secondary enlargement of the right heart occurs in all cases of long standing. In the arterio-sclerotic group there is an ever present possibility of narrowing of the orifices of the coronary arteries or an extension of the sclerosis to them, leading to fibroid myocardial degeneration. In the endocarditis cases the intima of the aorta may be perfectly smooth. The so-called dynamic dilatation of the arch is best seen in these cases. A young girl, whose case had been reported as one of aneurism, had forcible pulsation and a tumor which could be grasped above the sternum—post mortem the innominate artery did not admit the little finger and the arch was not dilated!

Although the coronary arteries are filled during systole, the circulation

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in them must be embarrassed as they must miss the effect of the blood pressure during the elastic recoil of the arteries, which surely aids in keeping the coronary vessels full. The arteries of the body usually present more or less sclerosis consequent upon the strain which they undergo during the forcible ventricular systole.

**Symptoms.**—The condition is often discovered accidentally in persons who have not presented any features of cardiac disease.

Headache, dizziness, and a feeling of faintness on rising quickly are among the earliest symptoms. Palpitation and distress on slight exertion are common. Long before any signs of failing compensation *pain* may be a marked feature. It is extremely variable in its manifestations. It may be of a dull, aching character confined to the præcordia but more frequently it is sharp and radiating, and transmitted up the neck and down the arms, particularly the left. Disease of the aorta is often responsible for pain. Angina pectoris is more frequent in this than in any other valve lesion. Anæmia is common, much more so than in aortic stenosis or mitral affections. Distressing dreams and disturbed sleep are more common in this than in other forms of valvular disease.

As compensation fails more serious symptoms are shortness of breath and œdema of the feet. The attacks of *dyspnoea* are liable to come on at night, and the patient has to sleep with his head high or even in a chair. Cyanosis is rare. It is most commonly due to complicating valve disease, or it is stated that it may result from bulging of the septum ventriculorum and encroachment upon the right ventricle. Of respiratory symptoms cough is common, due to the congestion of the lungs or œdema. Hæmoptysis is less frequent than in mitral disease but there are cases in which it is profuse and believed to be due to tuberculosis of the lungs. General dropsy is not common, but œdema of the feet may occur early and is sometimes due to the anæmia, sometimes to the venous stasis, at times to both. Unless there is coexisting mitral disease, it is rare for the patient to die with general anasarca. Sudden death is frequent; more so than in other valvular diseases. As compensation fails the patient takes to bed and irregular fever, associated usually with a recurring endocarditis, is not uncommon toward the close. Embolic symptoms are not infrequent—pain in the splenic region with enlargement of the organ, hæmaturia, and in some cases paralysis.

*Mental* symptoms are often seen with this lesion or the patients may be irritable and difficult to manage; toward the close there may be delirium, hallucinations, and morbid impulses. It is important to bear this in mind, for patients occasionally display suicidal tendencies.

**PHYSICAL SIGNS.**—*Inspection* shows a wide area of forcible impulse with the apex beat in the sixth or seventh interspace, and perhaps as far out as the anterior axillary line. In young subjects the præcordia may bulge. In very slight insufficiency there may be little or no enlargement. On *palpation* a thrill, diastolic in time, is occasionally felt, but is not common. The impulse is usually strong and heaving, unless in extreme dilatation, when it is wavy and indefinite. Occasionally two or three interspaces between the nipple line and sternum are depressed with systole as the result of atmospheric pressure. *Percussion* shows a great increase in the area of heart dullness, chiefly downward and to the left.

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*Auscultation.*—A diastolic murmur is heard at the base of the heart and propagated down the sternum. It may be feeble or inaudible at the aortic cartilage, and is usually heard best at midsternum opposite the third costal cartilage or along the left border of the sternum. It is usually blowing in quality, and is prolonged, or "long drawn," as the phrase is. It is produced by the reflux of blood into the ventricle. In some cases it is loudly transmitted to the axilla at the level of the fourth interspace, not by way of the apex. The second sound may be well heard or be replaced by the murmur, or with a dilated arch the second sound may have a ringing metallic or booming quality and the diastolic murmur is well heard, or even loudest, over the manubrium. Occasionally in the carotid artery the second sound is distinctly audible when absent at the aortic cartilage and, according to Broadbent, it is at the carotid that we must listen for the second aortic sound, for when heard it indicates that the regurgitation is small in amount, and is consequently a favorable prognostic element. In the larger arteries a systolic thud or shock may be heard and sometimes a double murmur.

The first sound may be clear at the base; more commonly there is a soft, short, systolic murmur. In the arterio-sclerotic group the systolic murmur is, as a rule, short and soft, while in the endocarditic group, in which the valve segments are united and often covered with calcified vegetations and excrescences, the systolic murmur is rough and may be accompanied by a thrill.

At the apex, or toward it, the diastolic murmur may be faintly heard propagated from the base. With full compensation the first sound is usually clear at the apex; with dilatation there is a loud systolic murmur of relative mitral insufficiency, which may disappear as the dilatation lessens.

*Flint Murmur.*—A second murmur at the apex, to which attention was called by the late Austin Flint, is not uncommon. It is of a rumbling, echoing character, occurring in the middle or latter part of diastole, and limited to the apex region. It is less intense than the murmur of mitral stenosis, and may be associated with a thrill. It is probably caused by the impinging of the regurgitant current from the aortic orifice on the large, anterior flap of the mitral valve, so as to cause interference with the entrance of blood at the time of auricular contraction. The condition is thus essentially the same as in a moderate mitral stenosis. This murmur is present in about half of the cases of uncomplicated aortic insufficiency (Thayer). It is very variable, disappearing and reappearing without apparent cause. The sharp, first sound and abrupt systolic shock, so common in true mitral stenosis, are rarely present, while the pulse is characteristic of aortic insufficiency.

*Arteries.*—The examination of the arteries is of great value. Visible pulsation is more commonly seen in the peripheral vessels in this than in any other condition. With the ophthalmoscope the retinal arteries are seen to pulsate. Not only is the pulsation evident, but the characteristic jerking quality is apparent. The throbbing carotids may lead to the diagnosis of aneurism. In many cases pulsation can be seen in the suprasternal notch and the abdominal aorta may lift the epigastrium with each systole. In severe cases with great hypertrophy, particularly if anæmia is present, the vascular throbbing may be of an extraordinary character, jarring the whole front of the chest, causing the head to nod, and even the tongue may throb rhythmically. To

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be mentioned with this is the *capillary pulse*, seen very often in aortic insufficiency, best in the finger nails or by drawing a line upon the forehead, when the margin of hyperæmia on either side alternately blushes and pales. In extreme grades the face or the hand may blush visibly at each systole. It is met with also in profound anæmia, occasionally in neurasthenia, and in conditions of great relaxation of the peripheral arteries. Pulsation may also be present in the peripheral veins. On palpation the characteristic *collapsing* or *Corrigan* pulse is felt. The pulse wave strikes the finger forcibly with a quick jerking impulse, and immediately recedes or collapses. This is sometimes best appreciated by grasping the arm at the wrist and holding it up. The pulse may be delayed—i. e., there is an appreciable interval between the beat of the heart and the pulsation in the radial artery, which varies according to the extent of the regurgitation.

The systolic pressure is variable and is often high; the diastolic is low and sometimes can not be obtained. This disproportion—a high pulse pressure—is characteristic. The systolic pressure in the femoral is higher than in the brachial artery. The sphygmographic tracing is very characteristic,



FIG. 13.—PULSE TRACING IN AORTIC INSUFFICIENCY; AN EXTRA SYSTOLE IS SHOWN.

showing the high ascent, the sharp top, and the quick drop in which the diastolic notch and wave are very slightly marked.

The studies of Stewart and of W. G. MacCallum showed that the low position of the diastolic notch in the descending arm of the pulse wave and the collapsing pulse are not due, as was formerly supposed, to the regurgitation in the left ventricle, but to dilatation of the peripheral arteries, which is a sort of protective adaptation under the vaso-motor influences.

Aortic insufficiency may be fully *compensated* for years and the patients may not suffer any inconvenience; the condition is often found accidentally. So long as the hypertrophy equalizes the valvular defect there may be no symptoms and the individual may take moderately heavy exercise without any distress. The cases which last the longest are those in which the insufficiency follows endocarditis and is not a part of a general arterio-sclerosis. The age at the time of onset is an important consideration, as in youth the lesion is not often from sclerosis, and the coronary arteries are unaffected. Coexistent lesions of the mitral valves tend to disturb compensation early. Pure aortic insufficiency is consistent with years of average health and with a tolerably active life. Alterations in the electro-cardiogram may aid in prognosis. Increase in the P R interval and the Q R S complex, inversion of the T wave in Lead II, and lack of correspondence in the routine findings are of serious import.

With the onset of myocardial changes, with increasing degeneration of the arteries, particularly with a progressive sclerosis of the arch and involvement of the orifices of the coronary arteries, the compensation becomes disturbed.

The insufficiency of the circulation is seen first on the arterial side in occasional fainting, giddiness, mental irritability and enfeeblement; later there may be mitral regurgitation and embarrassment of the right heart. In advanced cases the changes about the aortic ring may be associated with alterations in the cardiac nerves and ganglia and so introduce an important factor.

#### AORTIC STENOSIS

Definite aortic stenosis is a rare lesion. It may occur with insufficiency and probably in almost every case of stenosis there is some leakage.

**Etiology and Morbid Anatomy.**—In the milder grades there is adhesion between the segments, which are so stiffened that during systole they cannot be pressed back against the aortic wall. The process of cohesion between the segments may go on without great thickening, and produce a condition in which the orifice is guarded by a comparatively thin membrane, on the aortic face of which may be seen the primitive raphe separating the sinuses of Valsalva. In some instances this membrane is so thin and presents so few traces of sclerotic changes that the condition looks as if it was congenital. More commonly the valve segments are thickened and rigid with a cartilaginous hardness. In advanced cases they may be represented by calcified masses obstructing the orifice, through which a circular or slit-like passage is seen. The older the patient the more likely it is that the valves will be rigid and calcified.

We may speak of a *relative stenosis* when with normal valves and ring the aorta immediately beyond is greatly dilated. A stenosis due to involvement of the aortic ring in sclerotic changes without lesion of the valves is referred to by some authors but we have never met with an instance. A subvalvular stenosis, the result of endocarditis in the mitro-sigmoidean sinus, usually results from fetal endocarditis. Aortic stenosis is usually met with at a more advanced period of life than insufficiency, and the most typical cases are associated with extensive calcareous changes in the arterial system in old men.

Owing to the obstruction the ventricle works against increased resistance and its walls become hypertrophied, usually at first with little or no dilatation. In this condition are the most typical instances of concentric hypertrophy, in which, without much, if any, enlargement of the cavity, the walls are greatly thickened. The systole is prolonged, even as much as twenty-five per cent. There may be no changes in the other cardiac cavities if compensation is well maintained; but with its failure come dilatation, impeded auricular discharge, pulmonary congestion, and increased work for the right heart. The arterial changes are, as a rule, not so marked as in insufficiency. The amount of blood propelled through the narrow orifice may be smaller than normal, though when compensation is good the pulse wave may be of medium volume.

**Symptoms.**—The condition may be latent for an indefinite period, as long as the hypertrophy is maintained. Early symptoms are those due to defective blood supply to the brain, dizziness, and fainting. Palpitation, pain about the heart, and anginal symptoms are not so marked as in insufficiency. With myocardial failure, relative insufficiency of the mitral valve is established, and the patient presents the features of engorgement in the lesser and

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systemic circulations. Many of the cases in old people have symptoms pointing rather to general arterial disease. Cheyne-Stokes breathing is not uncommon with or without anæmia.

**PHYSICAL SIGNS.**—*Inspection* may fail to reveal any area of cardiac impulse, particularly in old men with rigid chest walls and emphysematous lungs. Under these circumstances there may be a high grade of hypertrophy without any visible impulse. Even when the apex beat is visible, it may be feeble and indefinite. In many cases the apex is displaced downward and outward, and the impulse looks strong and forcible.

*Palpation* reveals in many cases a *thrill* at the base of the heart of maximum force in the aortic region. With other condition do we meet with thrills of greater intensity. The apex beat may not be palpable or there may be a slow, heaving, forcible impulse.

*Percussion* never gives the same wide area of dulness as in aortic insufficiency and its extent depends largely on the state of the lungs.

*Auscultation.*—A rough systolic murmur is heard with maximum intensity at the aortic cartilage and propagated into the great vessels. One of the last lessons learned by the student is to recognize that a systolic murmur at the



FIG. 14.—PULSE TRACING IN AORTIC STENOSIS.

aortic area does not necessarily mean obstruction of the orifice. Roughening of the valves or of the intima of the aorta, and hæmic states are much more frequent causes. In aortic stenosis the murmur often has a much harsher quality, is louder and more frequently musical than in the conditions just mentioned. When compensation fails the murmur may be soft and distant. The second sound is rarely heard at the aortic area, owing to the thickening and stiffness of the valve. A diastolic murmur is not uncommon. Occasionally, as noted by W. H. Dickinson, there is a musical murmur of greatest intensity in the region of the apex, due probably to a slight regurgitation at high pressure through the mitral valves. The pulse in pure aortic stenosis is small, usually of good tension, well sustained, regular, and perhaps slower than normal.

**Diagnosis.**—With an extremely rough or musical systolic murmur of maximum intensity at the aortic region and transmitted to the vessels of the neck, hypertrophy of the left ventricle, a thrill, and a hard, slow pulse of moderate volume, which in a tracing gives a curve of slow rise, a broad, well sustained summit and slow decline, a diagnosis of aortic stenosis can be made with some degree of certainty, particularly if the subject is an old man. In aortic insufficiency a systolic murmur is usually present, but has neither the intensity nor the musical quality, nor is it accompanied by a thrill. With dilatation of the aorta, the murmur may be harsh or musical; but the existence of a sec-

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and sound, accentuated and ringing, is usually sufficient to differentiate this condition.

### MITRAL INSUFFICIENCY

**Etiology.**—Insufficiency of the mitral valve ensues: (a) From changes in the segments whereby they are contracted and shortened, usually combined with changes in the chordæ tendineæ, or with more or less narrowing of the orifice. (b) As a result of changes in the walls of the ventricle, either dilatation, so that the valve segments fail to close an enlarged orifice, or changes in the muscular substance, so that the segments are imperfectly coapted during the systole—*muscular insufficiency*. The common lesions producing insufficiency result from endocarditis, which causes a gradual thickening at the edges of the valves, contraction of the chordæ tendineæ, and union of the edges of the segments, so that in a majority of cases there is not only insufficiency, but some narrowing as well. Except in children, we rarely see the mitral leaflets curled and puckered without narrowing of the orifice. Calcareous plates at the base of the valve may prevent perfect closure of the segments.

In long-standing cases the entire mitral structures are converted into a firm calcareous ring. From valvular insufficiency the condition of muscular insufficiency must be carefully distinguished. It is met with in dilatation of the left ventricle, and also in weakening of the muscle in fevers and in anæmia.

**Morbid Anatomy.**—The effects of mitral insufficiency are as follows: (a) The imperfect closure allows blood to regurgitate from the ventricle into the auricle, so that at the end of auricular diastole this chamber contains not only the blood from the lungs, but also that regurgitated from the left ventricle. This necessitates dilatation, and, as increased work is thrown upon it in expelling the augmented contents, hypertrophy as well.

(b) With each systole of the left auricle a larger volume of blood is forced into the left ventricle, which dilates and subsequently hypertrophies.

(c) During the diastole of the left auricle, as blood is regurgitated into it from the left ventricle, the pulmonary veins are less readily emptied. In consequence the right ventricle expels its contents less freely, and in turn becomes hypertrophied and dilated.

(d) Finally, the right auricle also is involved, its chamber is enlarged, and its walls are increased in thickness.

(e) The effect upon the *pulmonary vessels* is to produce dilatation both of the arteries and veins—often in long-standing cases, atheromatous changes; the capillaries are distended, and ultimately brown induration is produced. Perfect compensation may be effected, chiefly through the hypertrophy of both ventricles, and the effect upon the peripheral circulation may not be manifested for years, as a normal volume of blood is discharged from the left heart at each systole. The time comes, however, when, owing either to increase in the grade of incompetency or to failure of compensation, the left ventricle is unable to send out its normal volume. Then there are overfilling of the left auricle, engorgement in the lesser circulation, embarrassed action of the right heart, and congestion in the systemic veins. For years this congested condition may be limited to the lesser circulation, but finally

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the tricuspid valves become incompetent, and the systemic veins are engorged. This leads to passive congestion and, when extreme, to œdema.

*Muscular insufficiency* is rarely followed by such perfect compensation. There may be in acute destruction of the aortic segments an acute dilatation of the left ventricle with relative mitral incompetency, great dilatation of the left auricle, and intense engorgement of the lungs, under which circumstances profuse hæmorrhage may result. In these cases there is little chance for the establishment of compensation. In cases of hypertrophy and dilatation of the heart, without valvular lesions, the insufficiency of the mitral valve may be extreme and lead to great pulmonary congestion, engorgement of the systemic veins, and cardiac dropsy, which can not be distinguished from that of mitral insufficiency due to lesion of the valve itself. In *chronic nephritis* the left ventricle may gradually fail, leading to relative mitral insufficiency and pulmonary and systemic congestion, similar to that induced by lesion of the valve itself. Adherent pericardium, especially in children, may lead to like results.

**Symptoms.**—During the development of the lesion, unless the insufficiency comes on acutely in consequence of rupture of the valve segment or of ulceration, the compensatory changes go hand in hand with the defect, and there are no subjective symptoms. So, also, in the stage of perfect compensation, there may be the most extreme grade of mitral insufficiency with enormous hypertrophy, yet the patient may not be aware of the existence of heart trouble, and may suffer no inconvenience except perhaps a little shortness of breath on exertion. It is only when the compensation has not been perfectly effected, or, having been so, is broken that the patients begin to be troubled. The symptoms may be divided into two groups:

(a) The *minor* manifestations while compensation is still good. Patients with extreme insufficiency often have a congested appearance of the face, the lips and ears have a bluish tint, and the venules on the cheeks may be enlarged—signs in many cases very suggestive. In long standing cases, particularly in children, the fingers may be clubbed, and there is shortness of breath on exertion. This is one of the most constant features and may exist for years, even when the compensation is perfect. Owing to the congested condition of the lungs these patients are liable to attacks of bronchitis or hæmoptysis. There may also be palpitation. As a rule, however, in well balanced lesions in adults, this period of full compensation is not associated with symptoms which call the attention to an affection of the heart, and with care the patient may reach old age in comparative comfort without being compelled to curtail seriously his pleasures or his work.

(b) Sooner or later comes a period of *broken compensation*, in which the most intense symptoms are those of venous engorgement. There are palpitation, weak, irregular action of the heart, and signs of dilatation. The irregularity may be due to extra-systoles or auricular fibrillation. Dyspnœa is an especial feature, and there may be cough. A distressing symptom is the cardiac "sleep-start," in which, just as the patient falls asleep, he wakes gasping and feeling as if the heart were stopping. There is usually slight cyanosis, and even slight jaundice. The most marked symptoms are those of venous stasis. The overfilling of the pulmonary vessels accounts in part for the dyspnœa. There is cough, often with bloody or watery expectoration, and the

alveolar epithelium containing brown pigment-grains is abundant. **Edema** usually sets in, beginning in the feet and extending to the body and the serous sacs. Right sided hydrothorax may recur and require repeated tapping. The urine is usually scanty and albuminous, and contains casts and sometimes blood cells. With judicious treatment compensation may be restored and all the serious symptoms pass away. Patients usually have recurring attacks of this kind, and die with a general dropsy; or there is progressive dilatation of the heart. Sudden death in these cases is rare. Some cases of mitral disease—stenosis and insufficiency—reach what may be called the *hepatic stage*, when all the symptoms are due to the secondary changes in the liver.

**PHYSICAL SIGNS.**—*Inspection.*—In children the præcordia may bulge and there may be a large area of visible pulsation. The apex beat is to the left of the nipple, in some cases in the sixth interspace, in the anterior axillary line. There may be a wavy impulse in the cervical veins, which are often full, particularly when the patient is recumbent.

*Palpation.*—The force of the impulse depends largely upon the stage. In full compensation it is forcible and heaving; when the compensation is disturbed, usually wavy and feeble.

*Percussion.*—The dulness is increased, particularly in a lateral direction. There is no disease of the valves which produces, in long standing cases, a more extensive transverse area of heart dulness. It does not extend so much upward along the left margin of the sternum as beyond the right margin and to the left of the nipple line.

*Auscultation.*—At the apex there is a systolic murmur which wholly or partly obliterates the first sound. It is loudest here, and has a blowing, sometimes musical character, particularly toward the latter part. It is transmitted to the axilla and may be heard at the back, in some instances over the entire chest. There are cases in which, as pointed out by Nauwyn, the murmur is heard best along the left border of the sternum. Usually at the apex the loudly transmitted second sound may be heard. Occasionally there is also a soft, sometimes a rough or rumbling presystolic murmur. As a rule, in cases of extreme mitral insufficiency from valvular lesion with great hypertrophy of both ventricles, there is heard only a loud blowing murmur during systole. A murmur of mitral insufficiency may vary a great deal, according to the position of the patient. An important sign is the accentuated pulmonary second sound.

The *pulse*, during the period of full compensation, may be full and regular, often of low tension. With the first onset of symptoms it may become irregular, a feature which then dominates the case throughout. Often after the disappearance of the symptoms of failure of compensation the irregularity persists. This is usually due to auricular fibrillation.

The three important physical signs of mitral regurgitation are: (a) Systolic murmur of maximum intensity at the apex, propagated to the axilla and heard at the angle of the scapula; (b) accentuation of the pulmonary second sound; (c) evidence of enlargement of the heart, particularly increase in the transverse diameter, due to hypertrophy of both ventricles.

**Diagnosis.**—There is rarely any difficulty in the diagnosis of mitral insufficiency. The physical signs are characteristic and distinctive. Two points are to be borne in mind. First, a murmur, systolic in time, and of maximum

intensity at the apex, and propagated even to the axilla, does not necessarily indicate mitral insufficiency. There is heard in this region a large group of what are termed accidental murmurs, the precise nature of which is doubtful. Some are cardio-respiratory.

Second, it is not always possible to say whether the insufficiency is due to lesion of the valve segment or to relative incompetency. The character of the murmur, the propagation, the accentuation of the pulmonary second sound, and the hypertrophy may not assist in the differentiation. The history is sometimes of greater value than the examination. The cases most likely to lead to error are those of the so-called idiopathic dilatation and hypertrophy (in which the systolic murmur may be of great intensity), and instances of arterio-sclerosis with dilated heart. Balfour and others maintain that organic disease of the mitral leaflets sufficient to produce insufficiency is always accompanied with some narrowing of the orifice, so that the only unequivocal proof of actual disease of the mitral valve is the presence of mitral stenosis.

#### MITRAL STENOSIS

**Etiology.**—There are two groups of cases, one following an acute endocarditis, the other the result of a slow sclerosis of the valves without any history of rheumatic fever or other infection. It is much more common in women than in men—in 4,791 autopsies at Guy's Hospital during ten years there were 196 cases, of which 107 were females and 89 males (Samways). This is not easy to explain, but there are at least two factors to be considered. Rheumatic fever prevails more in girls and chorea has an important influence, occurring more frequently in girls and being often associated with endocarditis. In a surprising number of cases of what the French call *pure* mitral stenosis no recognizable etiological factor can be discovered. This has been regarded by some as favoring the view that they may be of congenital origin, but congenital affections of the mitral valve are notoriously rare. Some suggest congenital syphilis. Whooping-cough, with its terrible strain on the heart, may be accountable for certain cases. While met with at all ages, stenosis is certainly most frequent in young adult women.

**Morbid Anatomy.**—The valve segments and chordæ may be fused together, the result of endocarditis. The condition varies a good deal, according to the amount of atheromatous change. In many cases the curtains are so welded together and the whole valvular region so thickened that the orifice is reduced to a mere chink—Orrigan's *button-hole contraction*. In non-endocarditic cases the curtains are not much thickened, but narrowing has resulted from gradual adhesion at the edges, and thickening of the chordæ tendineæ, so that from the auricle it looks cone like—the so-called *funnel-shaped variety*. The instances in which the valve segments are slightly deformed, but in which the orifice is considerably narrowed, are regarded by some as possibly of congenital origin. The involvement of the chordæ tendineæ is usually extreme, and the papillary muscles may be inserted directly upon the valve. In moderate grades of constriction the orifice admits the tip of the index finger; in more extreme forms the tip of the little finger; and occasionally one meets with a specimen in which the orifice seems almost obliterated. The heart is not greatly enlarged, rarely weighing more than

14 or 15 ounces. Occasionally, in an elderly person, it may seem only slightly, if at all, enlarged, and again there are instances in which the weight may reach as much as 20 ounces. The left ventricle is sometimes small, and may look very small in comparison with the right ventricle, which forms the greater portion of the apex. In cases in which with the narrowing there is insufficiency the left ventricle may be moderately dilated and hypertrophied.

It is not uncommon to find white thrombi in the appendix of the left auricle. Occasionally a large part of the auricle is occupied by an ante-mortem thrombus. Still more rarely the remarkable *ball thrombus* is found, in which a globular concretion, varying in size from a walnut to a small egg, lies free in the auricle.

The left auricle discharges its blood with greater difficulty and in consequence dilates, and its walls increase in thickness. Although the auricle is unfitted to compensate an extreme lesion, the probability is that for some time during the gradual production of stenosis the increasing power of the walls counterbalances the defect. In 36 cases of well-marked stenosis Samways found the auricle hypertrophied in 26, dilatation coexisting in 14. Eventually the tension is increased in the pulmonary circulation and extra work thrown on the right ventricle, which gradually hypertrophies. Relative incompetency of the tricuspid and congestion of the systemic veins supervene.

**Symptoms.**—Stenosis of the mitral valve may for years be efficiently compensated by the hypertrophy of the right ventricle. Many persons with this lesion present no symptoms. They may for years be short of breath on going upstairs, but carry on ordinary activity without discomfort. The pulse is smaller in volume than normal, and very often irregular (auricular fibrillation). A special danger is the recurring endocarditis. Vegetations may be whipped off into the circulation and, blocking a cerebral vessel, cause hemiplegia or aphasia, or both. This is not an uncommon sequence in women. Patients with mitral stenosis may survive this accident for an indefinite period.

**PHYSICAL SIGNS.**—*Inspection.*—There is often a flush on the cheeks, and clubbing of the fingers is common. The lower sternum and the fifth and sixth left costal cartilages are often prominent, owing to hypertrophy of the right ventricle. The apex beat may be ill defined. Usually it is not far beyond the nipple line, and the chief impulse is over the lower sternum and adjacent costal cartilages. Often in thin chested persons there is pulsation in the third and fourth left interspaces close to the sternum. When compensation fails, the impulse is much feebler, and in the veins of the neck there may be marked pulsation or the right jugular near the clavicle may stand out as a prominent tumor. In the later stage there is great enlargement with pulsation of the liver.

*Palpation* reveals in a majority of the cases a characteristic, well defined thrill, which is best felt, as a rule, at the apex or a little inside it. It is of a rough, grating quality, often peculiarly limited in area, most marked during expiration, and terminates in a sharp, sudden shock, synchronous with the impulse. This most characteristic of physical signs is pathognomonic of narrowing of the mitral orifice, and is perhaps the only instance in which the diagnosis of a valvular lesion can be made by palpation alone. It is often variable and may be brought out by exercise. The cardiac impulse is felt most forcibly over the lower sternum and in the fourth and fifth left inter-

spaces. The impulse is felt in the third and fourth interspaces, or in rare cases even in the second, and it has been thought that here it is due to pulsation of the auricle. It is always the impulse of the *conus arteriosus* of the right ventricle; even in the most extreme grades of mitral stenosis there is never such tilting forward of the auricle as would enable it to produce an impression on the chest wall.

*Percussion* gives an increase in dullness to the right of the sternum and along the left margin; not usually a great increase beyond the nipple line, except in extreme cases. There may be dullness in the left interscapular region.

*Auscultation.*—The findings are varied and puzzling combinations of sounds and murmurs may be heard. At the apex or a little inside it, often in a very limited region, there is heard a rough, vibratory or purring murmur, cumulative or crescendo in character, often of short duration, which terminates abruptly in the loud snapping first sound. This murmur is synchronous with the thrill and the loud shock with the first sound. The murmur may occupy the entire period of diastole, or the middle or only the latter half. A difference can often be noted between the earlier and later parts of the murmur, when it occupies the entire time. In some cases a soft diastolic murmur is heard after the second sound at the apex. This may increase and merge into the presystolic murmur. Often there is a peculiar rumbling or echoing quality, which in some instances is heard only over a single bell-space of the stethoscope. The administration of amyl nitrite may bring out the murmur more clearly. Some hold that the crescendo murmur is due to regurgitation and that the murmur occurs before the systolic sound but not before the contraction. It may persist with auricular fibrillation. In line with this, what is regarded as the true presystolic murmur is often faint, of low pitch and separated from the following sound or murmur. It is absent in auricular fibrillation; it may coexist with the crescendo murmur. A rumbling, echoing presystolic murmur at the apex is heard in some cases of aortic insufficiency (Flint murmur), occasionally in adherent pericardium with great dilatation of the heart, and in upward dislocation of the organ. The Graham Steell murmur of relative pulmonary insufficiency may be heard in the pulmonic area.

A systolic murmur may be heard at the apex or along the left sternal border, often of extreme softness and audible only when the breath is held. Sometimes the systolic murmur is loud and distinct and is transmitted to the axilla. The second sound in the second left interspace is loudly accentuated, and often reduplicated. It may be transmitted far to the left and be heard with great clearness beyond the apex. With good compensation the second sound is heard at the apex; its disappearance suggests the approach of decompensation. Other points to be noted are the following: The cause of the sharp, snapping first sound which follows the presystolic murmur is by no means easy to explain. It can scarcely be a valvular sound produced chiefly at the mitral orifice, since it may be heard with great intensity in cases in which the valves are rigid and calcified. It has been suggested that it is a loud "snap" of the tricuspid valves caused by the powerful contraction of the hypertrophied right ventricle. Broadbent thinks it may be due to the abrupt contraction of a partially filled left ventricle. The sound may be audible at

a distance, as one sits at the bedside of the patient (Graves). In one patient the first sound was audible six feet, by measurement, from the chest wall.

These signs are characteristic only of the stage in which compensation is maintained. The murmur may be soft, almost inaudible, and only brought out after exertion. Finally there comes a period in which, with the establishment of auricular fibrillation, the signs change. This is due to the absence of contraction of the auricle. Thus a presystolic murmur may disappear as there is not the usual difference in pressure in the auricle and ventricle at the time when the auricle should be contracting. With the auricle paralyzed the murmur is more likely to be heard early in diastole. Difference in rate may cause marked changes in the time and character of the murmur.

Sometimes in the apex region a sharp first sound or gallop rhythm may be heard. The systolic shock may be present after the disappearance of the thrill and the characteristic murmur. If partial heart-block occurs a complicated set of signs results as the auricle is contracting more often than the ventricle. With recovery of compensation and with increasing vigor of contraction of the right ventricle and left auricle, the presystolic murmur reappears. At this stage the nature of the valve lesion may be entirely overlooked. *Auricular fibrillation* is the rule in the arrhythmia of mitral stenosis.

Pressure of the enlarged auricle on the left recurrent laryngeal nerve, causing paralysis of the vocal cord on the corresponding side, has been described and the diagnosis of aneurism of the arch of the aorta may be made. Fetterolf and Norris conclude that it is not due to the pressure of the left auricle directly, but to squeezing of the nerve between the pulmonary artery and the aortic arch, and that the paralysis is due to the neuritis so excited.

Failure of compensation brings the group of symptoms discussed under cardiac insufficiency. Briefly enumerated, they are: Rapid and irregular action of the heart, shortness of breath, cough, signs of pulmonary engorgement, and very frequently hamoptysis. Attacks of this kind may recur for years. Bronchitis or a febrile attack may cause shortness of breath or cyanosis. Inflammatory affections of the lungs or pleura seriously disturb the right heart, and these patients stand pneumonia very badly. Many patients with mitral stenosis do not have dropsy. The liver may be greatly enlarged, and in the late stages ascites is not uncommon, particularly in children.

#### TRICUSPID VALVE DISEASE

**Tricuspid Regurgitation.**—This results from acute or chronic endocarditis very rarely. Nearly always the condition is one of relative insufficiency, and is secondary to lesions of the valves on the left side or myocardial insufficiency. It happens in a sound heart as a "safety valve" action when the pressure in the right heart is excessive, as in severe exertion. It is met with in conditions of the lungs which cause obstruction to the circulation, such as fibrosis and emphysema, particularly with chronic bronchitis. The symptoms are those of obstruction in the lesser circulation with venous congestion in the systemic veins. The signs are:

(a) Systolic regurgitation of the blood into the right auricle and the transmission of the pulse wave into the veins of the neck. If the regurgitation is slight or the contraction of the ventricle is feeble there may be no

venous throbbing, but in other cases there is marked systolic pulsation in the cervical veins, both in the internal and the external vein, particularly in the latter. Marked pulsation in these veins occurs only when the valves guarding them become incompetent. Slight oscillations are not uncommon, even when the valves are intact. The distention is sometimes enormous, particularly in the act of coughing, when the right jugular at the root of the neck may stand out, forming an extraordinarily prominent ovoid mass. Occasionally the regurgitant pulse wave may be widely transmitted and be seen in the subclavian and axillary veins, and even in the subcutaneous veins over the shoulder, or in the superficial mammary veins.

The regurgitant pulsation may be transmitted to the inferior cava, and so to the hepatic veins, causing pulsation of the liver. This is best appreciated by bimanual palpation. The pulsation may be readily distinguished, as a rule, from the impulse from the ventricle or transmitted from the aorta.

(b) The second important sign is the occurrence of a systolic murmur of maximum intensity over the lower sternum. It is usually a soft, low murmur, often distinguished from a coexisting mitral murmur by differences in quality and pitch, and may be heard to the right as far as the axilla. Sometimes it is very limited in its distribution. In addition, the percussion usually shows increase in dulness to the right of the sternum, and the impulse in the lower sternal region is forcible. In the great majority of cases the symptoms are those of the associated lesions. In fibrosis of the lung and chronic emphysema the failure of the right ventricle with tricuspid insufficiency may lead to gradual failure with cardiac dropsy.

**Tricuspid Stenosis.**—The condition is rare clinically and anatomically, and is not often recognized in life. Of 26,000 medical admissions in the Johns Hopkins Hospital there were only 8 with clinical or post mortem diagnosis of this condition; and in 3,500 autopsies, only 5 cases were found, all in females. Of 195 collected cases, there were 141 females, 38 males, 16 sex unknown. In a majority of the cases—104—the mitral and tricuspid were affected together, in 14 the tricuspid alone, in 64 the tricuspid and aortic. A definite history of rheumatism was present in only 66 cases (Futcher).

The *diagnosis* is not often made; extreme cyanosis and dyspnoea are common, and toward the end the ordinary signs of cardiac failure. Among the important physical signs are presystolic pulsation in the jugular veins and in the enlarged liver. A presystolic thrill may be felt at the tricuspid area with a marked systolic shock. The cardiac dulness is increased to the right, a rumbling presystolic murmur may be present over the lower sternum with an extension to the right border.

#### PULMONARY VALVE DISEASE

Murmurs in the region of the pulmonary valves are extremely common; lesions of the valves are exceedingly rare. Balfour has well called the pulmonary area the region of "auscultatory romance." A systolic murmur is heard here under many conditions—(1) very often in health, in thin-chested persons, particularly in children, during expiration and in the recumbent posture; (2) when the heart is acting rapidly, as in fever and after exertion; (3) it

is a favorite situation of the cardio-respiratory murmur; (4) in anæmic states; and (5) the systolic murmur of mitral insufficiency may be transmitted along the left sternal margin.

**Stenosis** is almost invariably a congenital anomaly and constitutes one of the most important of the congenital cardiac affections. The valve segments are usually united, leaving a small, narrow orifice. In adults cases occasionally occur. The congenital lesion is commonly associated with patency of the ductus Botalli and imperfection of the ventricular septum. There may also be tricuspid stenosis. Acute endocarditis not infrequently attacks the sclerotic valves. The physical *signs* are extremely uncertain. There may be a systolic murmur with a thrill best made out to the left of the sternum in the second intercostal space. This murmur may be like that of aortic stenosis, but is not transmitted into the vessels. The pulmonary second sound is weak or obliterated, or may be replaced by a diastolic murmur. Usually there is hypertrophy of the right heart.

**Pulmonary Insufficiency.**—This rare lesion was originally described by Morgagni. Pitt analysed 109 cases from the Guy's Hospital Reports, of which 60 had infectious endocarditis, 18 were due to a dilated pulmonary artery, 14 to pulmonary stenosis, 14 to aortic aneurism, 13 to abnormality in the number of the valves, and 6 unclassified. Pitt makes two groups, one with a rapid course, sometimes with definite symptoms pointing to the heart but the signs to general septicæmia. In the second group the cardiac symptoms are marked, dyspnœa, cough, etc., and the physical signs are definite.

The *signs* are those of regurgitation into the right ventricle, but, as a rule, it is difficult to differentiate the murmur from that of aortic insufficiency, though the maximum intensity may be in the pulmonary area. The absence of the vascular features of aortic insufficiency is important. Both Gibson and Graham Steell called attention to the possibility of leakage through these valves in cases of great increase of pressure in the pulmonary artery, and to a soft diastolic murmur heard under these circumstances.

#### COMBINED VALVULAR LESIONS

Valvular lesions are seldom single or pure; combined lesions are more common. This is particularly the case in congenital disease. In young children mitral and aortic lesions, the result of rheumatic fever, are common. Pure mitral insufficiency and pure mitral stenosis may exist for years, but in time the tricuspid becomes involved. Aortic valve lesions are more commonly uncombined than mitral lesions. The added lesion may be hurtful or helpful. The stenosis which so often accompanies the endocarditic variety may lessen the regurgitation in aortic insufficiency; and a narrowing of the mitral orifice may be beneficial in mitral regurgitation.

**Prognosis in Valvular Disease.**—The question is entirely one of efficient compensation. So long as this is maintained the patient may suffer no inconvenience, and even with the most serious forms of valve lesion the function of the heart may be little, if at all, disturbed. Practitioners should remember that the best judgment may be gathered from inspection and palpation. With an apex beat in the normal situation and a regular rhythm the auscultatory phenomena may be practically disregarded. *The myocardium is more impor-*

*tant than the valve.* A murmur *per se* is of little or no moment in determining the prognosis in any given case. There is a large group of patients who present only a systolic murmur over the body of the heart, or over the apex, in whom the left ventricle is not hypertrophied, the rhythm is normal, and who may not have had rheumatism.

Among the conditions influencing prognosis are: (a) AGE.—Children under ten are bad subjects. Compensation is well effected, and they are free from many influences which disturb compensation in adults. The coronary arteries are healthy and nutrition of the heart muscle can be readily maintained. Yet, in spite of this, the outlook in cardiac lesions in young children is usually bad. The valve lesion itself is apt to be progressive, and the limit of cardiac reserve force is early reached. There seems to be proportionately a greater degree of hypertrophy and dilatation. Among other factors of this period are insufficient food in the poorer classes, the recurrence of rheumatic attacks, and the existence of pericardial adhesions. The outlook in a child who can be carefully supervised and prevented from damaging himself by overexertion is better than in one who is constantly overtaking his circulation. The valvular lesions at, or subsequent to, puberty are more likely to be permanently and efficiently compensated. Sudden death from heart disease is very rare in children.

(b) SEX.—Women bear valve lesions, as a rule, better than men, owing partly to the fact that they live quieter lives, partly to the less common involvement of the coronary arteries, and to the greater frequency of mitral lesions. Pregnancy and parturition are disturbing factors, but are less serious than some writers would have us believe.

(c) VALVE AFFECTED.—The relative prognosis of the different valve lesions is difficult to estimate and each case must be judged on its own merits. *Aortic insufficiency* is unquestionably the most serious; yet for years it may be perfectly compensated. Favorable circumstances are a moderate grade of hypertrophy and dilatation and the absence of symptoms of cardiac distress, extensive arterio-sclerosis and angina. The prognosis rests largely with the condition of the coronary arteries. Rheumatic lesions of the valves, inducing insufficiency, are less apt to be associated with endarteritis at the root of the aorta; and in such cases the coronary arteries may escape for years. When aortic insufficiency is a part of an arterio-sclerosis at the root of the aorta, the coronary arteries are almost invariably involved, and the outlook is much more serious. This is often the case in syphilis. Sudden death is not uncommon, from acute dilatation during exertion, or, more frequently, from blocking of a coronary artery. The liability of this form to be associated with *angina pectoris* also adds to its severity. *Aortic stenosis* is comparatively rare, most common in middle aged or elderly men, and is, as a rule, well compensated.

In mitral lesions the outlook on the whole is more favorable than in aortic insufficiency. *Mitral insufficiency*, when well compensated, has a better prognosis than mitral stenosis. Except aortic stenosis, it is the only lesion commonly met with in patients over three-score years. The patients who last the longest are those in whom the valve orifice is more or less narrowed, as well as incompetent. There is no valve lesion so poorly compensated and so rapidly fatal as that in which the mitral segments are gradually curled and

puckered until they form a narrow strip around a wide mitral ring—a condition specially seen in children. There are cases of mitral insufficiency in which the defect is thoroughly balanced for thirty or even forty years, without distress or inconvenience. Even with great hypertrophy the compensation may be most effective. Women may pass safely through repeated pregnancies, though here they are liable to accidents associated with the severe strain.

In *mitral stenosis* the prognosis is usually regarded as less favorable but our experience places this lesion almost on a level, particularly in women, with mitral insufficiency. It is found very often in persons in perfect health, who have had no signs of heart-failure, and who have lived laborious lives. The figures given by Broadbent indicate that the date of death in mitral stenosis is comparatively advanced. Of 53 cases from the post mortem records of St. Mary's Hospital, thirty-three was the age for males, and thirty-seven or thirty-eight for females. These women pass through repeated pregnancies with safety. There is always the risk of cerebral embolism.

The outlook depends principally on the condition of the *myocardium*, which in large measure governs the situation. With evidence of muscular insufficiency the prognosis is always grave. The *etiological* factor is important, thus rheumatic fever or syphilis may have caused serious myocardial mischief. Every case must be judged separately, and all the circumstances carefully balanced. The development of auricular fibrillation, alternation of the heart, etc., must be taken into account. There is no question which requires greater experience and more mature judgment, and the most experienced are sometimes at fault. The following conditions justify a favorable prognosis: Good general health and good habits; no exceptional liability to rheumatic fever; origin of the valvular lesion independently of degeneration; existence of the valvular lesion without change for over three years; sound ventricles, of moderate frequency, and general regularity of action; the absence of serious forms of arrhythmia; sound arteries, with a normal tension; and freedom from pulmonary, hepatic, and renal congestion.

**Treatment.**—(a) STAGE OF COMPENSATION.—Medicinal treatment is not necessary and is often hurtful. A common error is to administer drugs, such as digitalis, on the discovery of a murmur or of hypertrophy. If the lesion has been found accidentally, it may be best not to tell the patient, but rather an intimate friend. Often it is necessary to be perfectly frank in order that the patient may take preventive measures. He should lead a quiet, regulated, orderly life, free from excitement and worry, and the risk of sudden death makes it imperative that the patient suffering from aortic disease should be warned against overexertion and hurry. An ordinary wholesome diet in moderate quantities should be taken; tobacco may be allowed in moderation, but alcohol should be interdicted or used in very small amount. Exercise should be regulated by the feelings of the patient. So long as no cardiac distress or palpitation follows, moderate exercise will prove beneficial. Any focus of infection should be treated. The skin should be kept active by a daily bath. Hot baths should be avoided and the Turkish bath forbidden. In the case of full-blooded, somewhat corpulent individuals, an occasional saline purge should be taken. Patients with valvular lesions should not go to very high altitudes. The act of coition has risks, particularly in aortic

disease. Knowing that the causes which most surely and powerfully disturb compensation are overexertion, mental worry, and malnutrition, the physician should give suitable instructions in each case. As it is always better to have the coöperation of an intelligent patient, he should, as a rule, be told of the condition, but in this matter the physician must be guided by circumstances, and there are cases in which reticence is the wiser policy.

(b) STAGE OF BROKEN COMPENSATION.—The break may be immediate and final, as when sudden death results from acute dilatation or from blocking of a branch of the coronary artery, or it may be gradual. Irregularity is not necessarily an indication of failing compensation but demands an accurate diagnosis. Serious failure of compensation is indicated by signs of dilatation, marked cyanosis, gallop rhythm, or certain forms of arrhythmia, with or without oedema. These are dependent on the myocardium and the same measures are indicated as given under the treatment of myocardial insufficiency. In mitral stenosis, valvulotomy has been done successfully (Cutler and Levine).

## V. SPECIAL PATHOLOGICAL CONDITIONS

### I. ANEURISM OF THE HEART

**Aneurism of a valve** results from acute endocarditis, which produces softening or erosion and may lead to perforation of the segment or to gradual dilatation of a limited area under the influence of the blood pressure. The aneurisms are usually spheroidal and project from the ventricular face of an aortic valve. They are much less common on the mitral segments. They frequently rupture and produce extensive destruction and insufficiency.

**Aneurism of the walls** results from the weakening due to chronic myocarditis, or occasionally follows acute mural endocarditis, which more commonly, however, leads to perforation. It has followed a stab wound, a gumma of the ventricle, and, according to some authors, pericardial adhesions. The left ventricle near the apex is usually the seat, because here fibrous degeneration is most common. Of 90 cases collected by Legg 59 were situated here. In the early stages the anterior wall of the ventricle, near the septum, sometimes even the septum itself, is slightly dilated, the endocardium opaque, and the muscular tissue sclerotic. In a more advanced stage the dilatation is pronounced and layers of thrombi occupy the sac. Ultimately a large rounded tumor may project from the ventricle and attain a size equal to that of the heart.

Occasionally the aneurism is sacculated and communicates with the ventricle through a small orifice. The sac may be double, as in the cases of Janeway and Sailer. In the museum of Guy's Hospital there is a specimen showing the wall of the ventricle covered with aneurismal bulgings. Rupture occurred in 7 of the cases collected by Legg.

The *symptoms* are indefinite. Occasionally there is marked bulging in the apex region and the tumor may perforate the chest wall. In mitral stenosis the right ventricle may bulge and produce a visible pulsating tumor below the left costal border, which has been mistaken for cardiac aneurism. When the sac is large and presses upon the heart, there may be a marked disproportion

between the strong cardiac impulse and the feeble pulsation in the peripheral arteries. The X-ray study may aid in diagnosis.

## II. RUPTURE OF THE HEART

This rare event is usually associated with fatty infiltration or myocardial degeneration. In some instances acute softening from embolism of a coronary artery, suppurative myocarditis, or a gummatous growth has been the cause. The majority of the patients are over sixty years of age. Schaps reports a case in an infant of four months associated with an embolic infarct of the left ventricle. Harvey, in his second letter to Riolan (1649), described the case of Sir Robert Darcy, who had distressing pain in the chest and syncopal attacks with suffocation, and finally cachexia and dropsy. Death occurred in one of the paroxysms. The wall of the left ventricle of the heart was ruptured, "having a rent in it of size sufficient to admit any of my fingers, although the wall itself appeared sufficiently thick and strong."

The rent may occur in any of the chambers, but is most frequent in the left ventricle on the anterior wall, not far from the septum. The accident usually takes place during exertion. There may be no preliminary symptoms, but without any warning the patient may fall and die in a few moments. Sudden death occurred in 71 of Quain's 100 cases. In other instances there may be a sense of anguish and suffocation, and life may be prolonged for several hours. In a Montreal case, the patient walked up a steep hill after the onset of the symptoms, and lived for thirteen hours. A case is on record in which the patient lived for eleven days.

## III. NEW GROWTHS AND PARASITES

Primary neoplasm is extremely rare. Secondary tumors may be single or multiple, and are usually unattended with symptoms, even when the disease is extensive. In one case in the right ventricle a mass was found which involved the anterior segment of the tricuspid valve and partly blocked the orifice. There were numerous cancerous emboli in the pulmonary artery. In another instance the heart was greatly enlarged, owing to the presence of innumerable masses of colloid cancer the size of cherries. Mediastinal sarcoma may penetrate the heart, though it is remarkable how extensive the disease of the mediastinal glands may be without involvement of the heart or vessels.

Cysts are rare. They are found in different parts, and are filled either with a brownish or a clear fluid. Blood cysts occasionally occur. Both the *Cysticercus cellulosæ* and echinococci cysts occur occasionally.

## IV. WOUNDS AND FOREIGN BODIES

Wounds of the heart may be caused by external injuries, as stabs and bullet wounds, by foreign bodies passing from the gullet or œsophagus, or by puncture for therapeutic purposes.

(a) Bullet wounds are common. Recovery may take place, and bullets may be encysted in the organ. Stab wounds are still more common. A med-

ical student, on a spree, passed a pin into his heart. The pericardium was opened, and the head of the pin was found outside of the right ventricle. It was grasped and an attempt made to remove it, but it was withdrawn into the heart and, it is said, caused the patient no further trouble (Moxon).

(b) Hysterical girls sometimes swallow pins and needles, which, passing through the œsophagus and stomach, are found in various parts of the body. A remarkable case is reported by Allen J. Smith of a girl from whom several dozen needles and pins were removed, chiefly from subcutaneous abscesses. Several years later she died of heart disease and needles were found in the tissues of the adherent pericardium, and between thirty and forty were embedded in the thickened pleural membranes of the left side.

(c) Puncture of the heart (cardiocentesis) has been done as a therapeutic procedure but is not without risk. Hæmorrhage may take place from the puncture, though it is not often extensive.

## VI. CONGENITAL AFFECTIONS OF THE HEART

These have only a limited clinical interest, as in a large proportion the anomaly is not compatible with life, and in others nothing can be done to remedy the defect or even to relieve the symptoms. Congenital affections result from interruption of the normal course of development or from inflammatory processes—endocarditis; sometimes from both.

**General Anomalies.**—Of general anomalies of development the following conditions may be mentioned: *Acardia*, absence of the heart, which has been met with in the monstrosity known by the same name; *double heart*, which has occasionally been found in extreme grades of fetal deformity; *dextrocardia*, in which the heart is on the right side, either alone or as part of a general transposition of the viscera; *ectopia cordis*, a condition associated with fission of the chest wall and of the abdomen. The heart may be situated in the cervical, pectoral, or abdominal regions. Except in the abdominal variety, the condition is very rarely compatible with extra-uterine life. Occasionally, as in a case reported by Holt, the child lives for some months, and the heart may be seen and felt beating beneath the skin in the epigastric region. This infant was five months old at the date of examination.

**Anomalies of the Cardiac Septa.**—The septa of both auricles and ventricles may be defective, in which case the heart consists of but two chambers, the *cor bilocularis* or reptilian heart. In the septum of the auricles there is a very common defect, owing to the fact that the membrane closing the foramen ovale has failed at one point to become attached to the ring, and leaves a valvular slit which may admit the handle of a scalpel. Neither this nor the small cribriform perforations of the membrane are of any significance.

The *foramen ovale* may be patent without a trace of membrane closing it. In some instances this exists with other serious defects, such as stenosis of the pulmonary artery, or imperfection of the ventricular septum. In others the patent foramen ovale is the only anomaly, and in many instances it does not appear to have caused any embarrassment, having been found in persons who have died of various affections. The ventricular septum may be absent, the condition known as *trilocular heart*. Much more frequently there

is a small defect in the upper portion of the septum, either in the situation of the membranous portion known as the "undefended space" or in the region just anterior to this. This is frequently associated with narrowing of the pulmonary orifice or of the conus arteriosus of the right ventricle.

Apart from the instances in association with narrowing of the orifice of the pulmonary artery, or of the conus, there are cases in which defect of the *membranous septum* is the only lesion, a condition not incompatible with long and fairly active life. The late Professor Brooks of the Johns Hopkins University knew from early manhood that he had heart trouble, but he accomplished an extraordinary amount of work, and lived to be about 60. Imperfect septum was the only lesion. The physical signs are fairly distinctive, with usually some evident enlargement of the heart, and a murmur described by Roger as follows: "It is a loud murmur, audible over a large area, and, commencing with systole, is prolonged so as to cover the normal tic-tac. It has its maximum, not at the base to the right, as in aortic stenosis, or to the left, as in pulmonary stenosis, but at the upper third of the præcordial region. It is central, like the septum, and from this central point gradually diminishes in intensity in every direction. The murmur does not vary at any time, and it is not conducted into the vessels." In some cases there is a distinct systolic intensification of this loud continuous murmur.

*Anomalies and Lesions of the Valves.*—Numerical anomalies of the valves are not uncommon. The semilunar segments at the arterial orifices are not infrequently increased or diminished in number. Supernumerary segments are more frequent in the pulmonary artery than in the aorta. Four, or sometimes five, valves have been found. The segments may be of equal size, but, as a rule, the supernumerary valve is small.

Instead of three there may be only two semilunar valves, or, as it is termed, the *bicuspid condition*: this is more frequent in the aortic valve. Of 21 instances only 2 occurred at the pulmonary orifice. Two of the valves have united, and from the ventricular face show either no trace of division or else a slight depression indicating where the union has occurred. From the aortic side there is usually to be seen some trace of division into two sinuses of Valsalva. There has been a discussion as to the origin of this condition, whether it is really an anomaly or due to endocarditis, fetal or post-natal. The combined segment is usually thickened, but the fact that this anomaly is met with in the fetus without a trace of sclerosis or endocarditis shows that it may, in some cases at least, result from a developmental error.

Clinically this is a very important congenital defect, owing to the liability of the combined valve to sclerotic changes. Except two fetal specimens, all of a series showed thickening and deformity, and in 15 of those reported death resulted directly or indirectly from the lesion (Osler).

The little fenestrations at the margins of the sigmoid valves have no significance; they occur in a considerable proportion of all bodies.

Anomalies of the auriculo-ventricular valves are not often met with.

FETAL ENDOCARDITIS may occur either at the arterial or auriculo-ventricular orifices. It is nearly always of the chronic or sclerotic variety. Very rarely, indeed, is it of the warty or verrucose form. There are little nodular bodies, sometimes six or eight in number, on the mitral and tricuspid segments—the nodules of Albini—which represent the remains of fetal structures,

and must not be mistaken for endocardial outgrowths. The little rounded, bead-like hæmorrhages of a deep purple color, which are very common on the heart valves of children, are also not to be mistaken for the products of endocarditis. In fetal endocarditis the segments are usually thickened at the edges, shrunken, and smooth. In the mitral and tricuspid valves the cusps are found united and the chordæ tendineæ are thickened and shortened. In the semilunar valves all trace of the segments has disappeared, leaving a stiff membranous diaphragm perforated by an oval or rounded orifice. It is sometimes difficult to say whether this has resulted from fetal endocarditis or is an error in development. In many instances the processes are combined; an anomalous valve becomes the seat of sclerotic changes. According to Rauchfuss, endocarditis is more common on the right side of the heart only because the valves are more often the seat of developmental errors.

**LESIONS AT THE PULMONARY ORIFICE.**—Stenosis is one of the commonest and most important of congenital heart affections. A slow endocarditis causes gradual union of the segments and narrowing of the orifice to such a degree that it admits only the smallest sized probe. In some of the cases the smooth membranous condition of the combined segments is such that it suggests faulty development. In some instances vegetations occur. The condition is compatible with life for many years, and in a considerable proportion of the cases of congenital heart disease above the tenth year this lesion is present. With it there may be defect of the ventricular septum. Pulmonary tuberculosis is a common cause of death. Obliteration or *atresia* of the pulmonary orifice is a less frequent but more serious condition than stenosis. It is associated with persistence of the ductus arteriosus, together with patency of the foramen ovale or defect of the ventricular septum with hypertrophy of the right heart. *Stenosis of the conus arteriosus* of the right ventricle exists in a considerable proportion of the cases of pulmonary stenosis. At the outset a developmental error, it may be combined with sclerotic changes. The ventricular septum is imperfect, the foramen ovale usually open, and the ductus arteriosus patent. The lesions at the pulmonary orifice constitute the most important group of congenital cardiac affections. Of 631 instances of congenital anomalies analyzed by Maude Abbott, 150 cases came under this category.

**CONGENITAL LESIONS OF THE AORTIC ORIFICE** are not very infrequent. Rauchfuss collected 24 cases of stenosis and *atresia*; stenosis of the left conus arteriosus may also occur, a condition not incompatible with prolonged life. Ten of the 16 cases tabulated by Dilg were over thirty years of age.

**TRANSPOSITION OF THE LARGE ARTERIAL TRUNKS** is a not uncommon anomaly. There may be neither hypertrophy, cyanosis, nor heart murmur.

**Symptoms of Congenital Heart Disease.**—Cyanosis occurs in over 90 per cent. of the cases, and forms so distinctive a feature that the terms "blue disease" and "*morbus cæruleus*" are practically synonyms for congenital heart-disease. The lividity in a majority of cases appears only within the first week of life, and may be general or confined to the lips, nose, and ears, and to the fingers and toes. In some instances there is in addition a general dusky suffusion, and in the most extreme grades the skin is almost purple. It may vary a good deal and be intense only on exertion. The external temperature is low. *Dyspnœa* on exertion and *cough* are common symptoms. A great

increase in the number of the red corpuscles is common, and they may reach 8 or 9 million. There may be nucleated red cells and great variation in size and shape. The children rarely thrive, and often display a lethargy of both mind and body. The fingers and toes are *clubbed* to a degree rarely met with in any other affection. The cause of the cyanosis has been much discussed. Morgagni referred it to the general congestion of the venous system due to obstruction. Morrison's analysis of 75 cases of congenital heart disease shows that closure of the pulmonary orifice with patency of the foramen ovale and ventricular septum is the condition most frequently associated with cyanosis, and he concludes that the deficient aëration of the blood owing to diminished lung function is the most important factor. Another view, often attributed erroneously to William Hunter, was that the discoloration was due to the admixture in the heart of venous and arterial blood; but lesions may permit of very free mixture without producing cyanosis. The question of the cause of cyanosis can not be considered as settled. Variot made the suggestion that the cause is not entirely cardiac, but is associated with disturbance throughout the whole circulatory system, and particularly a vaso-motor paresis and malaëration of the red blood cells.

**Diagnosis.**—In children, cyanosis, with or without enlargement of the heart, and the existence of a murmur, are sufficient, as a rule, to determine the presence of a congenital heart lesion. The cyanosis gives us no clew to the precise nature of the trouble, as it is common to many lesions and may be absent in certain conditions. The murmur is usually systolic. It is not always present, and there are instances of complicated congenital lesions in which the examination showed normal heart sounds. In two or three instances fetal endocarditis has been diagnosed *in gravis* by the presence of a rough systolic murmur, and corroborated subsequent to the birth of the child. Hypertrophy is present in a majority of the cases of congenital defect. The fatal event may be caused by abscess of the brain. For a full discussion of the subject the student is referred to the monograph of Dr. Maude Abbott in our System of Medicine.

The conclusions of Hochsinger are as follows:

“(1) In childhood, loud, rough, musical heart murmurs, with normal or only slight increase in the heart dulness, occur only in congenital heart disease. The acquired endocardial defects with loud heart murmurs in young children are almost always associated with great increase in the heart dulness. In the transposition of the large arterial trunks there may be no cyanosis, no heart murmur, and an absence of hypertrophy.

“(2) In young children heart murmurs with great increase in the cardiac dulness and feeble apex beat suggest congenital changes. The increased dulness is chiefly of the right heart, whereas the left is only slightly altered. On the other hand, in the acquired endocarditis in children, the left heart is chiefly affected and the apex beat is visible; the dilatation of the right heart comes late and does not materially change the increased strength of the apex beat.

“(3) The entire absence of murmurs at the apex, with their evident presence in the region of the auricles and over the pulmonary orifice, is always an important element in differential diagnosis, and points rather to septum defect or pulmonary stenosis than to endocarditis.

"(4) An abnormally weak second pulmonic sound associated with a distinct systolic murmur is a symptom which in early childhood is only to be explained by the assumption of a congenital pulmonary stenosis.

"(5) Absence of a palpable thrill, despite loud murmurs which are heard over the whole præcordial region, is rare except with congenital defects in the septum, and it speaks, therefore, against an acquired cardiac affection.

"(6) Loud, especially vibratory systolic murmurs, with the point of maximum intensity over the upper third of the sternum, associated with a lack of marked symptoms of hypertrophy of the left ventricle, are very important for the diagnosis of a persistence of the ductus Botalli, and can not be explained by the assumption of an endocarditis of the aortic valve."

Escherich suggests that the systolic basic murmur heard sometimes in the newborn, particularly if premature, may originate in the ductus Botalli before its closure.

**Treatment.**—The child should be warmly clad and guarded from all circumstances liable to excite bronchitis. In the attacks of urgent dyspnoea with lividity venesection is advisable. Saline cathartics are useful. Digitalis must be used with care; it is sometimes beneficial in the later stages. When compensation fails, the indications for treatment are those of muscular insufficiency in acquired cardiac disease.

## VII. ANGINA PECTORIS

**Definition.**—A symptom-complex characterized by paroxysmal attacks of pain, usually pectoral, associated with vascular changes.

**History.**—In 1768 Heberden described a "disorder of the breast," to which he gave the name of "Angina Pectoris." Before this date Morgagni and Rougnon had described cases. The association with coronary artery disease was early shown by Jenner. John Hunter died in an attack. The connection with aortitis as demonstrated by Corrigan and Allbutt, the recognition of extra-pectoral forms, and the introduction of nitrites in treatment by Lauder Brunton are the important contributions of the nineteenth century.

**Etiology.**—The disease is not uncommon; there were 1,090 deaths in England and Wales in 1922. In the United States the death rate is increasing; there were 7,571 deaths in the registration area in 1920.

It is a rare disease in the public wards of hospitals. It is a disease of the better classes, and a consultant in active work may see a dozen or more cases a year.

**AGE.**—In our series of 300 cases there were, under 30, 9 cases; between 30 and 40, 42; between 40 and 50, 60; between 50 and 60, 93; between 60 and 70, 72; between 70 and 80, 20; above 80, 4.

**SEX.**—Of our cases 256 were men and 44 women.

**RACE.**—The disease seems to be relatively more frequent in the United States. Jews are particularly prone (42 of our 300 cases).

**OCCUPATION.**—It is not an affection of the working classes. The life of stress and strain, particularly of worry, seems to predispose to it, and this is perhaps why it is so common in our profession. In our series of 300 cases there were 38 physicians, a large proportion. From John Hunter onward a

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long list of distinguished physicians have been its victims, among whom in recent years were Charcot, Nothnagel, and William Pepper.

**CARDIO-VASCULAR DISEASE.**—In persons under forty syphilis is an important feature, causing an aortitis, often limited to the root of the vessel. Whatever the cause, arterio-sclerosis predisposes to angina. A majority of the patients have sclerosis, many high blood pressure. Business and professional men leading lives of great strain, and eating, drinking, and smoking to excess, form a large proportion of angina cases. Slight attacks may occur with high blood pressure alone.

**HEREDITY.**—The disease may occur in members of three generations, as in the Arnold family.

**Imitative Features.**—Outbreaks of angina-like attacks have been described. After the death of one member of a family from the disease, another may have somewhat similar attacks. Two of his physicians had angina after Senator Sumner's fatal attack. One of them died within two weeks; the other, a young man, recovered completely.

**Pathology.**—The lesions in 17 post mortems were as follows:

(a) *Coronary artery disease* was present in 13 cases. The orifices only may be involved in a sclerotic aortitis. In one case they were narrowed to admit only a bristle, while the vessels beyond were normal. Blocking of a branch with a fresh thrombus, or with an embolus may occur. During an attack an infarct may soften, with perforation of the ventricular wall. Obliterative endarteritis was present in 9 of the cases. In elderly subjects the coronary vessels may be calcified—the condition found by Jenner in John Hunter.

(b) *Aortitis* was present in four of the cases, in syphilitic subjects, all under 40 years of age. Corrigan first called attention to this lesion in angina, the great importance of which has been emphasized by Clifford Allbutt. It is usually limited to the supra-sigmoidal area.

(c) In a few instances no lesions have been found. In one case of the list a man aged 26 had attacks, which were regarded as functional, on and off for two years. Death occurred after a series of paroxysms. The aorta was small, otherwise there were no changes.

**Pathogenesis.**—No generally accepted explanation of the phenomena of the attack has been offered and probably various factors may be responsible. It has been regarded as a neuralgia of the cardiac nerves, a cramp of the heart muscle, or of parts of it, a manifestation of myocardial ischaemia, or an expression of tension of the ventricular walls. The intermittent claudication theory of Allan Burns may be defined as a state in which an artery admits enough blood to a muscular structure for quiet work, but not enough for increased work, so that the contractile function of the muscle is disturbed and pain results. Sir James Mackenzie regards the disease as an expression of cardiac exhaustion and also of a susceptible nervous system. The pain is a viscerosensory reflex: the feeling of constriction a visceromotor reflex. Reid suggests that failure of reflex dilatation of the peripheral vascular system may cause a sudden rise of pressure in the aorta and left ventricle. The increased tension irritates the nerve supply.

We owe much to Sir Clifford Allbutt for his studies on the disease, which should be consulted by every student. His explanation is that in the large

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proportion of cases (say 90 per cent.) the attacks are due to disease of the thoracic aorta, especially in its outer coat where there are sensory end-organs regulating blood-pressure. Tension of the first portion of the aorta is an important factor. The coronary arteries and myocardium have little or nothing to do with the pain but much with the mortality. Sudden death is due to vagus inhibition. There is strong evidence that this explanation is the most satisfactory one. Very similar attacks of pain occur in acute and in some cases of chronic aortitis. In many cases aortic pain can be clearly distinguished from cardiac pain; some patients have both and recognize the difference. The picture produced by embolism of the coronary arteries can usually be distinguished from angina pectoris; the symptoms due to coronary artery disease otherwise are practically those of myocardial degeneration. It is well to realize that the origin of severe thoracic pain, due to disease of the circulatory system, may be difficult to determine.

A number of conditions may be responsible for an attack. (1) *Physical exertion* in almost any form. Walking on the level *slowly*, the patient may be comfortable but increase beyond a certain pace or going up a slight grade brings on an attack. Some patients are affected by walking against a wind or fast driving in an open motor car. Any sudden exertion or hurrying does the same. (2) *Emotion and excitement*. These are important causes, anger or annoyance being specially likely to cause an attack. (3) *Cold*. This may be shown on general chilling or by going out on a cold day, on taking a cold bath or even on washing the face in cold water. (4) *Digestive disturbance*, especially with distention after a heavy meal, is a common cause. Numerous other causes may operate, such as stooping over, straining at stool, coitus, etc. In nearly all there is the influence of strain and influences acting on the peripheral circulation.

Disease of the heart muscle is not a cause and the features of this are not present in angina pectoris. The onset of myocardial failure alters the picture and with it the features of angina disappear in the majority of cases. The localization of the pain is that of disease of the aorta and not of the myocardium.

**Symptoms.**—Classified by the severity of the attacks, cases may be grouped in three categories:

(a) **MILDEST FORM** (*“Les Formes Frustes”* of the French).—There is a feeling of substernal tension, uneasiness, or distress, rising at times to positive pain, usually associated with emotion, sometimes with exertion, but soon passing off. There may be slight pallor, or feeling of faintness. When rising to speak in public there may be a feeling of substernal tension—it is a common experience—which passes off. Muscular effort, as in climbing a hill or a stair, may bring on the sensation. In the high pressure life a man may experience for weeks or months this sense of substernal tension, not pain, and without accurate localization or radiation, and not increased by exercise or emotion. It is as, one patient expressed it, a “hot-box” indicating too great pressure and too high speed. It is away after the night’s rest, and may disappear entirely when the “harness” is taken off.

(b) **MILD FORM** (*Angina Minor*).—Pain of moderate severity with radiation to the arm is met with in nervous persons, in tobacco smokers, sometimes following the acute infections, particularly influenza. The attacks are

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brought on by emotion, more frequent in women, and never fatal. Often called pseudo, false, functional, or toxic angina, the difference in the character of the attacks may be one of degree only. The conditions under which the attacks come on are of greater importance than the nature of the attack itself. There may be marked vaso-motor disturbance, with cold, numb, and blue extremities, followed by præcordial pain and a feeling of faintness. In persons addicted to tea, coffee, and tobacco heart pain is not infrequent, sharp and shooting, associated with palpitation, or severe and truly anginal.

(c) SEVERE ANGINA (*Angina Major*).—The two special features in this group are the existence in a large proportion of all the cases of organic disease of heart or vessels and the liability to sudden death. An exciting cause of the attack can usually be traced. John Hunter used to say that "his life was in the hands of any rascal who chose to worry him," and his fatal attack occurred in a fit of anger.

PHENOMENA OF THE ATTACK.—The patient is seized with agonizing pain, usually about the upper sternum, and a sense of constriction, sometimes of extreme degree. The pain may begin at the lower end of the sternum and travel upwards. The pain usually radiates to the left shoulder and down the left arm. It may radiate to the left back, to the neck, angle of the jaw or to the scalp. The extent of referred pain varies greatly. It may radiate down both arms or to the right arm only. There may be numbness of the fingers or in the cardiac region. The face is usually pallid and may assume an ashy gray tint, and not infrequently there is profuse sweating. The paroxysm lasts from several seconds to a minute or two, during which, in severe attacks, the patient feels as if death were imminent. As pointed out by Latham, there are two elements in it, the pain—*dolor pectoris*—and the indescribable feeling of anguish and sense of imminent dissolution—*angor animi*. There are great distress and anxiety, and the patient may drop dead at the height of the attack or faint and pass away in syncope. The condition of the heart during the attack is variable; the pulsations may be uniform and regular. The pulse tension, however, is usually increased, but it is surprising, even in cases of extreme severity, how slightly the character of the pulse may be altered. After the attack there may be eructations or the passage of a large quantity of clear urine. The patient usually feels exhausted, and for a day or two may be badly shaken; in other instances in an hour or two he feels himself again. While dyspnoea is not a constant feature, it may occur with the paroxysm; there may be bronchial wheezing which may come on rapidly with dyspnoea.

Death may occur in the first attack, as in the well known case of Thomas Arnold; or at the end of a series of attacks, the so-called *status anginosus*. Paroxysms may occur for a year or more before the fatal attack.

There is a *chronic* form in which attacks occur irregularly. John Hunter's first seizure was in 1773, and he had many in the 20 years before his death. Sometimes life is a terrible burden, as any emotion or effort may bring on an attack. And, lastly, after paroxysms of great severity recurring for months, or even for so long as two years, complete recovery takes place.

EXTRA-PECTORAL FEATURES.—In the attack the pain usually radiates up the neck and down the left arm. As the studies of Mackenzie and Head have shown in disease of the heart and of the aorta, the pain is referred to the

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1st, 2d, 3d, and 4th dorsal areas; and in angina it may be also in areas of the distribution of the 5th to the 9th dorsal nerves. The pain may begin in the left arm, or in the jaw, even in the front teeth, or in one testis. Sometimes the pain remains in these distant parts, and yet the attack presents, as noted by Heberden, all the features of angina. The attack may begin with agonizing pain in the left leg or in the left pectoral muscle. The entire features of the attack may be subdiaphragmatic—the so-called *angina abdominalis*. In at least twelve of our series the pains were abdominal and various diagnoses may be made.

The *pulmonary features* are remarkable. A condition like acute emphysema may come on, with wheezing and an inflated state of the lungs. Acute oedema may follow with the expectoration of large quantities of a thin, bloody fluid. The blood pressure may be extraordinarily high—340 mm. Hg. in one case. *Cerebral features* are not common, but unconsciousness, transient monoplegia, or hemiplegia and aphasia may occur.

**Diagnosis.**—In typical severe attacks there is rarely any difficulty, especially if an attack is observed or a good description is obtained. In all cases careful inquiry should be made as to the situation and radiation of the pain, as well as its character. The pain of angina is referred to the upper sternum in the majority of cases; pain felt about the apex or over the lower precordium is probably not anginal. The sense of constriction, the attitude and color of the patient, and sweating are important. In acute and chronic *aortitis* attacks occur which can hardly be distinguished but as a rule they are not of maximum severity and last longer than the usual attack of angina. In coronary thrombosis or embolism, the onset is sudden and without apparent cause, the signs of shock or collapse are more marked, the heart action is very weak, the pulse is rapid and small, the blood pressure usually drops to low figures, there may be leucocytosis, the pain persists for a longer period (unless death comes quickly) and may not be relieved by nitrites or morphia. The diagnosis of *abdominal angina* may be possible only if an attack is observed. In cases of less severe pain, particularly if not localized, the diagnosis may be very difficult. Careful inquiry should be made as to causal factors, as the influence of exertion and emotion is important, with thorough study for other causes of pain. It is a good rule to treat doubtful cases as if they were angina. The occurrence of attacks of moderate or slight severity must be remembered.

**Prognosis.**—In men under 40 syphilis must be suspected, and with appropriate treatment recovery may be complete (see the Lumleian Lectures (Osler), *Lancet*, 1910, 1). In men in the 5th and 6th decades who have lived the high pressure life a change of habits may bring relief; but, as Walshe remarked, "the cardinal fact in real angina is its uncertainty." Even after attacks of the greatest severity recovery is possible. The circumstances that bring on an attack are important. The angina that follows any slight exertion is, as a rule, more serious than that which comes on spontaneously or is excited by emotion; yet one patient who could never dress without having what he called "angor de toilette" lived for 11 years. A rapid fall in blood pressure is of grave significance. The electrocardiographic study is of value as abnormal tracings are important. Irregularity or spreading of the Q R S complex in all leads and inversion of the T wave are regarded as serious indica-

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tions. The cardio-vascular condition is of the first importance in prognosis. Very high blood pressure, advanced arterio-sclerosis, marked disease of the aorta, valvular disease and signs of myocardial weakness are of serious import. A large proportion of all cases have no obvious signs of cardiac disease; the coronary arteries may be extensively diseased with clear heart sounds and a good pulse. In women the forms of angina with marked vaso-motor disturbance as a rule do well, and when neurotic or hysterical manifestations are prominent the outlook is good.

There are three modes of dying in angina—one, as Walshe says, "is sudden, instantaneous, coeval with a single pang." The functions of life stop abruptly, and with a gasp all is over. Ventricular fibrillation may be the cause. In a second mode, following a series of attacks, the heart grows weaker and the patient dies in a progressive asthenia; while in a third there is a gradually induced cardiac insufficiency with dyspnoea.

**Treatment.**—Prolonged rest is important and the general treatment should be much the same as for aneurism. Every effort should be made to reduce anxiety and sources of irritation. Factors which induce an attack should be avoided. The diet should be simple and the bowels kept freely open. If there are any signs of myocardial insufficiency, even without loss of compensation, an occasional course of digitalis is advisable. Syphilitic cases require active treatment—neoarsphenamine in small doses followed by mercury and iodide of potassium. In the neurotic cases with a recognition of the basic disturbance in the vaso-motor system a rest cure and hydrotherapy are indicated. Ergotin grs. ii (0.13 gm.) three times a day has a definite value in vaso-motor instability. It is always well to give iodide in the dosage found best for the patient. The effect of vaso-dilators should be tried and the one used which is most useful. Many patients do well by taking sodium nitrite (gr. i, 0.06 gm. three times a day) regularly and carrying nitroglycerine with them so that it can be taken at once (gr. 1/100-1/50) on any feeling of discomfort or pain. The taking of nitroglycerine and morphine at the first sign of an attack is often successful in preventing severe manifestations. The use of theobromine (gr. xv, 1 gm.) or diuretin (gr. xv, 1 gm.) three times a day is sometimes of benefit. In the severer types the treatment is concerned with the attack and with the general condition afterward. In the attack inhalation of nitrite of amyl, introduced by Lauder Brunton, may give instant relief. We see its benefit particularly in cases with widespread arterial constriction. In the recurring terrible paroxysms it may lose its effect, but many patients are relieved promptly, and it gives great comfort and confidence to carry the *perles*. Morphia should be used freely when amyl nitrite fails and when the attacks recur with great frequency. As Burney Yeo pointed out, angina patients are very resistant to this drug. Chloroform may have to be used, and is always helpful, never harmful. With a dusky cyanosis and asthma-like breathing oxygen inhalations may be given.

**Surgical.**—A number of procedures have been carried out, the object being to interrupt the connection between the aorta and nerve centres and so prevent irritation of the cardio-aortic plexus. Jonnesco removes the left cervical sympathetic chain including the superior middle and inferior cervical ganglia and the superior thoracic ganglion. Division of the left superior cardiac branch of the cervical sympathetic and the main trunk below the ganglion

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has been successful (Brown and Coffey) and also division of the depressor nerve. In some cases there is no doubt that "the danger signal" is removed but operation holds out hope for some patients.

### C. DISEASES OF THE ARTERIES

#### I. ACUTE ARTERITIS

This is a rare condition which occurs most often with acute infectious disease, especially typhoid fever. In the aorta it may follow an infectious endocarditis or infection may occur through the vasa vasorum in septicæmia or pyæmia, or spread from bronchial lymph-nodes or the pericardium. It may be associated with bacterial (mycotic) aneurisms.

*Symptoms.*—If the process is in the aorta, any symptoms are part of the general process and can not be separated. If in the peripheral arteries the onset is usually with severe pain in the vessel, sometimes with redness and swelling. The pulse is obliterated in the artery below the affected spot and the limb becomes cold and white. The results depend on the collateral circulation very largely; gangrene does not always follow. In *treatment* little can be done beyond keeping the part thoroughly warm and relieving pain.

#### II. ARTERIO-SCLEROSIS

The conception of arterio-sclerosis as an independent affection—a general disease of the vascular system—is due to Gull and Sutton. Arterio-sclerosis in one sense is not a disease but comprises changes which may occur with or follow many diseases. The term designates anatomical changes rather than a clinical condition in itself. The clinical features result mainly from interference with the blood supply of organs or tissues, and the resulting disturbance of function. Any classification of its forms must be morphological and not clinical; the effort to separate too many forms causes confusion.

*Definition.*—A vascular lesion characterized by pathological thickening of the arterial walls, due probably to various factors, with an inflammatory reaction and degenerative changes as important elements, while hyperplastic and involutionary processes play a part; no one process is entirely responsible. In the large vessels the changes are often designated as atheroma; in the smallest vessels as obliterative endarteritis.

*Etiology.*—Among the important factors are the following:

(a) **HYPERTENSION.**—The blood pressure depends upon five factors: The heart pump supplies the force; the elastic coats of the large arteries store and convert an intermittent into a continuous stream; the small arteries act as sluices or taps regulating the control to different parts; the capillary bed is the irrigation field over which the nutritive fluid is distributed; and the drainage system is represented by the veins and lymph channels.

Galen first grasped the fact that life depends upon the maintenance of a due pressure in these irrigation fields: "Many canals dispersed throughout all

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the parts of the body convey to them blood as those of a garden convey moisture, and the intervals separating those canals are wonderfully disposed by nature in such a way that they should neither lack a sufficient quantity of blood for absorption, nor be overloaded at any time with an excessive supply."

The blood pressure varies greatly in different individuals and in the same individual under varying conditions. The normal pressure is from 120 to 130 mm. of mercury, but in persons over 50 it is often from 130 to 150 mm. A permanent pressure above 160 mm. may be called high, but there are great regional variations.

The relation of hypertension to arterio-sclerosis has been much discussed. Briefly, there are three groups of cases: (1) First, the simple high tension without signs of arterial or renal disease—what Clifford Allbutt calls *hyperpiesia*. In this condition, met with in individuals otherwise healthy, the blood pressure is permanently high—above 180—but, so far as can be ascertained, there are no arterial, cardiac, or renal changes. It is difficult to exclude internal, not discernible alterations in the splanchnic and other vessels, since vascular disease may be very localized. But clinically the group is well defined and very important. The tendency is to advance to the second group unless the hypertension disappears. The condition is met with most frequently in keen business men, who work hard, drink hard, and smoke hard.

The exact cause of this high tension we do not know. Some have attributed it with lack of evidence, to over-activity of the adrenals, but it is much more likely that the primary difficulty is somewhere in the capillary bed—in that short space in which the real business of life is transacted. Probably *diffuse hyperplastic sclerosis* is often responsible, beginning first in the smaller vessels of the kidney and spleen. However produced, the important point is that this hypertension leads to arterio-sclerosis, which can be produced experimentally by the injection of epinephrine and other hypersensitive substances.

(2) In the second group of cases the high tension is associated with an arterio-sclerosis with consecutive cardiac and renal disease.

(3) In the third group the high tension is secondary to forms of chronic nephritis in association with cardio-vascular disease.

(b) As an INVOLUTION PROCESS arterio-sclerosis is natural with old age, and is the expression of the natural wear and tear to which the tubes are subjected. Longevity is a vascular question, which has been well expressed in the axiom that "a man is only as old as his arteries." To a majority of men death comes primarily or secondarily through this portal. The onset of what may be called physiological arterio-sclerosis depends, firstly upon the quality of arterial tissue (vital rubber) which the individual has inherited, and secondly upon the amount of wear and tear to which he has subjected it. That the former plays a most important rôle is shown in the cases in which arterio-sclerosis sets in early in life in individuals in whom none of the recognized etiological factors can be found. Thus a man of thirty may have the arteries of a man of sixty, and a man of forty may present vessels as much degenerated as at eighty. Entire families sometimes show this tendency to early arterio-sclerosis, which can not be explained in any other way than that in the makeup of the machine bad material was used for the tubing. The influence of heredity plays a considerable part but more commonly the arterio-sclerosis results from the bad use of good vessels.

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(c) **INTOXICATIONS.**—Alcohol, lead, and gout play an important rôle in the etiology, although the precise mode of their action is not clear. They may act, as Traube suggests, by increasing the peripheral resistance in the smaller vessels and so raising the blood pressure, or possibly, as Bright taught, they alter the quality of the blood and render more difficult its passage through the capillaries. The toxins of disturbed metabolism or of the acute infections may produce degenerative changes in the media and adventitia. Thayer called attention to the frequency of arterial changes as a sequence of typhoid fever.

(d) **SYPHILIS** is one of the most important single causes.

(e) **OVEREATING.**—This plays an important part in inducing arterio-sclerosis. George Cheyne's advice is always applicable.

(f) **STRESS AND STRAIN.**—There are men in the fifth decade who have not had syphilis or gout, who have eaten and drunk with discretion, and in whom none of the ordinary factors are present—men in whom the arterio-sclerosis seems to come on as a direct result of a high pressure life.

(g) **OVERWORK OF THE MUSCLES**, which acts by increasing the peripheral resistance and by raising the blood pressure.

(h) **RENAL DISEASE.**—The relation between the arterial and kidney lesions has been much discussed. There are two groups of cases, one in which the arterio-sclerosis is primary, and the other in which it is secondary to a primary affection of the kidneys.

**Morbid Anatomy.**—The affection is met with most frequently in the aorta and its main branches. It is comparatively less frequent in the mesenteric and rare in the pulmonary arteries. Several forms may be recognized:

(a) **NODULAR.**—The aorta presents in the early stages numerous flat projections, yellowish or yellowish white in color, and situated particularly about the orifices of the branches. The initial change is in the intima, probably due to bacterial toxins, and the lesions in many ways resemble those of the next group. In the early stage these patches are scattered and do not involve the entire intima. In more advanced stages the patches undergo atheromatous changes. The material undergoes softening and breaks up into granular material, consisting of molecular debris—the so-called atheromatous abscess. Klotz has called attention to the frequency of nodular endarteritis about the orifices of the intercostal arteries in young people, usually in association with acute infections.

(b) **DIFFUSE ARTERIO-SCLEROSIS.**—In this form, the “arterio-capillary fibrosis” of Gull and Sutton, well termed diffuse hyperplastic sclerosis, met with usually in middle-aged men, or younger persons, the affection is widespread throughout the arteries. The essential lesion is a thickening of the intima in the smaller arteries, which is uniform in the circumference. In the arterioles the first change is swelling and proliferation of the endothelial cells, which undergo hyaline swelling with the production of fatty granules. Gradually the cell outline is lost and the protoplasm replaced by fat. The nuclei disappear. The fatty degeneration stops where the arteriole joins the parent vessels, in which the intima is thickened with the same hyaline change, and there may be great increase in the elastic tissue. The changes in the endothelium are an essential part of the process; the hyaline change may be inflammatory or degenerative. Hypertrophy of the media occurs more in

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the larger vessels. There is much difference of opinion as to the relative part played by inflammatory and degenerative processes. There is much to be said for the view that bacterial toxins play a large part in the etiology (see Evans, Goulstonian Lectures, *Brit. Med. Jour.*, March, 1923). In this group the heart hypertrophies and fibrous myocarditis is often present. The kidneys are sclerotic, may be increased in size, and are usually firm. In places the surface may be rough, or present atrophied depressed areas of a deep red color.

(c) SENILE ARTERIO-SCLEROSIS.—In this the larger arteries are dilated and tortuous, the walls thin and stiff, and the smaller vessels, as the radials, converted into rigid tubes like pipe-stems. The intima of the aorta may be occupied by rough, calcareous plaques, with here and there fissures and loss of substance. There may be sub-endothelial softening with the formation of atheromatous ulcers on which thrombi may deposit; though there may be extreme calcification and roughness with erosions of the aorta without thrombus formation. In the smaller vessels, as the radials, there are degeneration and calcification of the media—the so-called Monckeberg type.

(d) SYPHILITIC ARTERIO-SCLEROSIS.—In the aorta this is usually a mesoaortitis with definite characteristics. Macroscopically it may be limited in extent, localized at the root of the aorta, or about the orifice of an aneurism, or there is a band of an inch in width on some portion of the tube, while other parts of the aorta and its branches are normal. In other instances the intima is involved, not with the usual plaque-like areas of atheroma, but there are shallow depressions of a bluish tint and short transverse or longitudinal puckerings sometimes with a stellate arrangement; or the intima is pitted and scarred with depressions and linear sulci. Microscopically the most important changes are in the media and adventitia: (1) perivascular infiltration of the vasa vasorum; (2) small-celled infiltration in areas of the media, with (3) splitting, separation, and destruction of elastic fibres and the muscle cells. The intima over these areas may be perfectly normal, but often shows signs of thickening with fatty degeneration and the production of hyaline. Similar changes have been described by Klotz in the larger blood-vessels in cases of congenital syphilis. The specific nature of this mesoaortitis has been determined by the detection of the spirochaetes. Other forms affecting the smaller vessels have been referred to under syphilis.

(e) SCLEROSIS OF THE PULMONARY ARTERY is met with in various conditions: (1) With high tension, particularly in emphysema and mitral disease, the sclerosis may be marked, the main branches may be dilated, and the valves thickened and incompetent. (2) Gummatous arteritis has been met with (Warthin). (3) Primary sclerosis is not uncommon in India (Leonard Rogers). Aneurismal dilatation may be present. Syphilis is a factor in some cases in which dyspnoea, cyanosis, polycythemia, repeated hæmoptysis, angina with enlargement of the heart and chronic passive congestion are features. Our South American colleagues call it "Ayerza's disease."

In many cases of arterio-sclerosis the condition is not confined to the arteries, but extends not only to the capillaries but also to the veins, and may properly be termed an *angio-sclerosis*.

(f) SCLEROSIS OF THE VEINS—*phlebo-sclerosis*—is not an uncommon accompaniment of arterio-sclerosis. It is seen in heightened blood pressure, as in the portal system in cirrhosis of the liver and in the pulmonary veins in

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mitral stenosis. The affected vessels are usually dilated, and the intima shows a compensatory thickening, which is particularly marked in those regions in which the media is thinned. The new formed tissue in the endophlebitis may undergo hyaline degeneration and is sometimes extensively calcified. Without arterio-sclerosis the peripheral veins may be sclerotic, usually in conditions of debility, but not infrequently in young persons.

**SYMPTOMS.**—The early symptoms are interesting. Stengel has called attention to the pallor, and there may be dyspeptic symptoms. It is remarkable with what rapidity the disease may progress. The peripheral arteries may stiffen and grow old in a couple of years.

The combination of hypertension, palpable thickening of the arteries, hypertrophy of the left ventricle, and accentuation of the aortic second sound is common in one form of arterio-sclerosis. From the period of establishment the course may be very varied. For years the patient may have good health, and be in a condition analogous to that of a person with a well-compensated valvular lesion. There may be no renal symptoms, or there may be the passage of a larger amount of urine than normal, with transient albuminuria, and now and then hyaline casts. The subsequent history is extraordinarily diverse, depending upon the vascular territory in which the sclerosis is most advanced, or upon the accidents which are so liable to happen, and the symptoms may be cardiac, cerebral, renal, etc. In some cases there is a rapid loss of weight.

(a) *Cardiac.*—Involvement of the coronary arteries may lead to various symptoms—thrombosis with sudden death, fibroid degeneration of the heart, aneurism of the heart and rupture. An important group of symptoms results from the dilatation which finally gets the better of the hypertrophy. The patient presents all the symptoms of cardiac insufficiency and when he comes under observation for the first time the clinical picture may be that of chronic valvular disease, and a loud blowing murmur at the apex may throw the practitioner off his guard. Many cases terminate in this way.

(b) *Blood Pressure.*—This depends upon the degree of peripheral resistance and the force of the ventricular contraction. A high pressure pulse may exist with very little arterio-sclerosis; but, as a rule, when the hypertension has been persistent, sclerosis and high tension are found together. On the other hand, a low or normal pressure may be present in extremely sclerotic vessels.

(c) The *cerebral* symptoms are varied and embrace those of many degenerative diseases, acute and chronic (which follow sclerosis of the smaller branches), and cerebral hæmorrhage. Syphilis should always be considered in these cases. Transient hemiplegia, monoplegia, or aphasia may occur in advanced arterio-sclerosis. The attacks are characteristic, often brief, lasting twenty-four hours or less. Recovery may be perfect. Recurrence is the rule, and a patient may have a score or more attacks of aphasia, or in a couple of years there may be half a dozen transient hemiplegic attacks or one or two monoplegias, or paraplegia for a day or two. These cases seem best explained on the view of transient spasm. Vertigo occurs frequently, and may be simple or associated with slow pulse and syncopal or epileptiform attacks—the Stokes-Adams syndrome. The cerebral features are discussed more fully on page 1012.

(d) *Renal* symptoms supervene in a large number of cases. A sclerosis,

patchy or diffuse, is present in a majority of the cases at autopsy, and the condition is practically that of contracted kidney. It is seen typically in the senile form, and not infrequently develops early in life as a direct sequence of the diffuse variety. It is often difficult to decide clinically (and the question is one upon which good observers might not agree in a given case) whether the arterial or the renal disease has been primary.

(e) *Abdominal Arterio-sclerosis*.—It is believed to be associated particularly with overeating and chronic overtaking of the stomach and intestines. The condition is not uncommon, and the sclerosis of the splanchnic vessels may be advanced out of all proportion to that elsewhere. The *symptoms* are indefinite, sometimes resembling those of the ordinary neurosis with marked constipation, features that are by no means certainly associated with sclerosis; on the other hand, there is much more reason to connect the attacks of severe abdominal pain, the gastric crises of lead and of tabes with spasm of the vessels in this condition. There are cases of angina pectoris with abdominal pain which may be due to angiospasm of the sclerotic vessels.

(f) *Gangrene* of the extremities may be due directly to endarteritis or to dislodgment of thrombi. Sudden transient paralysis of the legs may occur.

(g) *Sclerosis of the Vessels of the Legs*.—The main symptom is pain in the legs, after walking for a few minutes or on walking fast, which may pull the patient "up short" or gradually reach a point at which motion is impossible. The patient rarely falls and after resting for a few minutes he can again walk. The attacks are similar to those of angina pectoris; as one intelligent man expressed it—"there is no difference in the sensation, it is only in the place." Cramp of the muscles may occur, and aggravate the pain, sometimes in paroxysms of severe intensity, or nocturnal cramp may be troublesome. Numbness, tingling and sensations of cold are common, and when dependent the feet may become deeply congested. Routine examination of the arteries of the foot should be made; endarteritis obliterans may be suggested by this vaso-motor disturbance. The arteries of the feet may be felt as hard cords without pulsation and phlebo-sclerosis is common.

*Intermittent lameness* or *claudication*, the dysbasia angio-sclerotica of Erb, the crural angina of Walton, is associated with arterio-sclerosis. In the horse, in which the intermittent lameness was first described by Bouley, verminous aneurisms are present in the aorta or the iliac arteries. In man, Charcot described the condition in 1856 in an old soldier who was not able to walk for more than a quarter of an hour without severe cramps in the legs. The post mortem showed a traumatic aneurism of one iliac artery. The loss of function and the pain in the muscles are due to the relative ischæmia. Of 127 cases there were only 7 in women (Erb). Hebrews seem more frequently affected. Syphilis, alcohol, and tobacco are common factors.

*Thrombo-angiitis obliterans* (Buerger) is an acute inflammatory lesion with occlusion thrombosis, probably due to infection. There is excruciating pain in the foot and leg, worse at night. The feet are blue and congested, and the skin clammy with decreased sensitiveness to heat and cold. There may be atrophy of the toes with dark colored skin and sometimes gangrene. Pulsation in the vessels of the affected leg is decreased or absent.

**Diagnosis.**—The recognition of thickening of the arterial walls is usually easy but care should always be taken to feel the radial artery when empty of

## DISEASES OF THE CIRCULATORY SYSTEM

blood. The temporal artery is often prominent in thin persons without being much thickened. Palpation should always include several arteries in addition to the radial—the brachial, the temporal, the femoral and the arteries of the foot. Search should be made for alteration in the aorta (dilatation) and the retinal arteries should be inspected. The diagnosis should include the blood pressure, the state of the kidneys and the heart. Diagnosis of the form of arterio-sclerosis is possible to some extent. Calcareous deposit may be felt. In the cases with hypertension and renal changes, the sclerosis is usually of the diffuse hyperplastic form.

**Prognosis.**—This depends greatly on the renal and myocardial condition. With advanced disease in either or both the ultimate outlook is grave. The distribution of the vascular changes is not uniform and signs of special involvement in one organ are more serious than thickening of the peripheral vessels. In rapidly advancing cases the outlook is grave. If the process is stationary and due to some past disease, such as typhoid fever, there is little change in the life expectation if the myocardium and kidneys are not damaged.

**Treatment.**—In the late stages the conditions must be treated as they arise in the various viscera. In the early stages, before any local symptoms are manifest, the patient should be enjoined to live a quiet well regulated life, avoiding excesses in food and drink. Ordinary exercise is beneficial and the patient should not limit his physical activity unless there are other reasons for it. If the patient is overweight this should be reduced gradually. The diet should be of simple food, quantity being more important than quality. A moderate amount of protein should be taken as it is usually harmful to reduce this too much. The intake of salt should be kept down; a sufficient amount is added in cooking without using it at table. It is usually best to explain frankly the condition of affairs, and so gain his intelligent coöperation. Special attention should be paid to the bowels and urine, and the skin should be kept active by daily baths. Alcohol in all forms should be prohibited. The use of mineral waters or a residence every year at one of the mineral springs is usually serviceable. If there has been a syphilitic history the persistent use of iodide of potassium is indicated; indeed, even in the non-syphilitic cases it seems to do good. It is best given in small doses, grains v to x (0.3 to 0.6 gm.). Whenever the blood pressure is high nitroglycerine or sodium nitrite may be given to relieve symptoms rather than with any hope of essentially influencing the disease. For intermittent claudication the operation of peri-arterial sympathectomy is sometimes very successful. In thrombo-angiitis obliterans W. A. Steel advises keeping the patient in bed for a month with the leg in an electric light hot air bath at about 110°. Every second day 250 c. c. of a 2 per cent. sodium citrate solution is given intravenously. After a month the sodium citrate is given every third or fourth day and the patient is allowed in a wheel chair with the leg down for short periods. Later the injections are given less often and the activity gradually increased. Iodide (gr. 10, 0.6 gm.) is given thrice daily throughout.

In cases which come under observation for the first time with dyspnoea, slight lividity, and signs of cardiac insufficiency, venesection is indicated. In some instances, with very high tension, striking relief is afforded by the ab-

straction of 10 to 20 ounces of blood. Cardiac failure, renal symptoms, etc., require the usual treatment.

### III. ALTERATIONS IN BLOOD PRESSURE

Variations in blood pressure do not constitute diseases but result from disturbances in many diseased conditions. The arterial pressure depends on (1) the power of contraction of the left ventricle, (2) the volume of blood forced into the aorta, (3) the condition of the arteries, especially their elasticity, (4) the vaso-motor tone, particularly in the capillaries, (5) the peripheral resistance and (6) the viscosity of the blood. The blood pressure is not fixed in health but varies especially with exercise and nervous disturbance.

**Hypertension.**—To warrant this term, the increase in pressure must be permanent, and 160 may be taken as an arbitrary figure for the minimum of hypertension. *Etiology.*—Clinically it occurs in a variety of conditions, always remembering that there may be a combination of factors. The most important are: (1) *Hypertrophy* of the left ventricle. (2) *Essential hypertension, hyperpiesia.* The cause of this is obscure but it often leads to changes in the vessels and to renal changes, which may terminate in chronic nephritis with hypertension. This sequence of events is proved; the mechanism of the process is doubtful in many cases. (3) *With renal disease.* The maximum figures are found in this group, especially in chronic interstitial nephritis. Some are primarily renal, others secondary to general arterial disease. (4) With some cases of *arterio-sclerosis*, probably especially in those termed diffuse hyperplastic sclerosis. (5) With *endocrine* gland disturbance. This includes many puzzling cases. Hypertension in women about the menopause is comparatively common. In some cases it may last for some months or a year and then disappear. (6) With *intracranial* conditions causing increased intracranial pressure, such as hæmorrhage. (7) With *increased volume of blood.* (8) With increased viscosity of the blood. (9) With *obesity.*

*Pathogenesis.*—This is obscure in many cases and there are probably several factors. (1) *Mechanical.* These operate in hypertrophy of the ventricle and in the nephritic-sclerotic group. (2) *Nervous.* The effect is apparently through the vaso-motor system and possibly by influences on the endocrine glands. (3) *Chemical and toxic.* Of these we know very little and it is here perhaps that much of the etiology lies. The view that excessive secretion of epinephrine is often responsible has little to support it. What part bacterial toxins play is a question but low-grade long continued infections may be important. Major finds decreased excretion of guanidine bases.

*Symptoms.*—These are variable and difficult to separate from those due to other conditions. In many cases there are none; the individual is in good health and the hypertension is discovered accidentally. Headache, vertigo, feelings of fullness in the head and flushing are common complaints. What may be termed "hypertension hypochondriasis" is frequent. The patients suffer more from knowing that hypertension is present than from the hypertension itself. They study their blood-pressure figures as some do those of the stock market. The physical findings may show only high pressure and in temporary cases there may be nothing more. Usually changes in the

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arteries are evident, or appear later, with cardiac hypertrophy and renal changes. In many cases the hypertension is only part of the picture. The *diagnosis* offers no difficulty but should not be made on one reading unless there are definite cardiac, renal, or cerebral changes.

*Prognosis.*—This depends principally on the cause and the extent of disease in the circulatory and renal systems; to a less extent on the ability of the patient to follow the best method of life.

*Treatment.*—The etiology should be studied, any causal factors removed if possible and other lesions properly treated. Any focus of infection should be removed. How much should the patient be told as to the hypertension and the figures? Good judgment is needed to decide this, but it is rarely wise to frighten him with talk of hæmorrhage, etc. A frank explanation of the condition and the reasons for the treatment advised is usually wise, with the advice not to have the pressure taken frequently. Often the question arises as to how much reduction in work is advisable. Each patient is a special problem but in general it is wise to advise cutting down the amount of business or work, especially if it involves worry and strain. Mental rest and quiet, so far as they can be secured, are important. Long hours of physical rest are useful. Exercise, short of fatigue, is helpful, best in the form of walking, golf, etc. A good vacation, often well spent at one of the springs, is an advantage. One day a week in bed on a low diet is useful. The *diet* should not be too much restricted, as the mistake of too much reduction in protein is often made. The quantity of food should be reduced in the majority. Meat, poultry or fish may be taken once a day in moderate amounts. Cereals, vegetables and fruits can be used; eggs need not be forbidden. Tea and coffee are to be used in moderation; alcohol should not be taken. Water should be taken fairly freely, especially before meals. It is well to reduce the amount of salt; none should be added to the food. If the patient is over-weight, by proper diet and exercise, this should be reduced. In such patients it is well to reduce the carbohydrate intake.

Bathing in tepid or warm water is usually best. The bowels should be kept well open, for which a saline before breakfast is often useful. A weekly dose of blue mass (gr. 5-10, 0.3-0.6 gm.) or mercury and chalk powder (gr. 5, 0.3 gm.) at bedtime for two successive nights is often beneficial. Some patients do well with irrigations of the colon once or twice a week in addition. The use of *vaso-dilators* is sometimes of value in relieving symptoms, especially headache and dizziness. Nitroglycerine, sodium nitrite and erythrol tetranitrate are the most useful. The dose must be decided for each patient. Any effort to reduce the pressure by drugs is useless and inadvisable, except in the patients with endocrine disturbance. Here no rules can be given; in the cases at the menopause, the giving of ovarian or corpus luteum extract may be effectual. Iodide is rarely of great value.

*Hypotension.*—A systolic figure below 110 may be regarded as hypotension.

*Etiology.*—(1) Weakness of the left ventricle. (2) *Vaso-motor disturbance*, especially in some infections, as typhoid fever, and in general nervous debility. (3) In some cases of *arterio-sclerosis*. (4) In *anæmia*. (5) In conditions in which the amount of blood is decreased as after hæmorrhage or severe diarrhœa. (6) In some *endocrine gland disturbances*. Special interest

## AORTITIS

belongs to the question of *adrenal insufficiency* which may be a factor in some cases. (7) With *cachexia*.

*Symptoms.*—It is difficult to separate these from associated conditions. Headache and dizziness are common, especially on getting up suddenly from a recumbent position or on stooping. Weakness is common and fatigue is easily produced. Syncope may occur frequently. Special interest attaches to the *white line* produced by gently drawing the nail over the skin of the body. It appears slowly and may persist for some minutes. Some regard it as diagnostic of adrenal insufficiency; the invariable association is very doubtful. Vaso-motor instability is often marked. As a rule there is a less proportionate fall in the diastolic than in the systolic and pulse pressures. The prognosis depends on the cause; some have a low pressure naturally and are in perfect health.

*Treatment.*—This may be clearly etiological as in myocarditis or anaemia. In general every effort should be made to improve the general health. Exercise has to be carefully ordered as these patients are easily fatigued; simple exercises which can be done with the patient lying down are useful. Rest should be ample and an hour or two of rest in the day is an advantage. Cool baths or sponging with brisk rubbing is helpful. Drugs sometimes help but often have no effect. Ergot, as the fluid extract (5 ss, 2 c. c.) or ergotin (gr. ii, 0.13 gm.), is often useful. Pituitary extract (gr. ii, 0.13 gm.) is worth a trial. Adrenal gland therapy is best given as the whole gland extract (gr. 5, 0.3 gm.). Digitalis is not indicated except for myocardial insufficiency.

## IV. AORTITIS

**Acute Aortitis.**—This is much more common than is usually recognized. It occurs in the acute infections but most especially in septicæmia and rheumatic fever, particularly in children who have aortic endocarditis. Of great importance is its occurrence in syphilis.

*PATHOLOGY.*—The process may be diffuse or most evident in slightly raised areas which at first are soft and later harder and with a yellow tinge. The first portion of the arch is most often affected and this may involve the orifices of the coronary arteries. If the aorta was previously diseased, all stages of atheroma may be found.

*SYMPTOMS.*—Pain is common, usually referred to the upper part of the sternum and sometimes radiating into the arms. There may be dyspnoea and a sense of thoracic oppression. In the syphilitic form, pain is the outstanding symptom, sometimes with the characters of angina pectoris. In other forms the pain is merged in the symptoms of the primary condition, especially in the acute infections.

*SIGNS.*—There may be marked pulsation in the neck, especially in the suprasternal notch, where the aorta may be seen and felt, and in the first and second interspaces. There is dulness over the manubrium and in the first two interspaces, both to right and left. The second sound may have a musical bell-like quality, sometimes very characteristic. The *syphilitic* form as a rule shows in addition the signs of aortic insufficiency.

*DIAGNOSIS.*—The main requisite is that the condition be kept in mind. It

is unrecognized because not considered. If there is any doubt the X-ray examination should decide. A positive Wassermann reaction or other evidence of syphilis gives the diagnosis of this form.

**PROGNOSIS.**—The condition in itself probably does not shorten life but may lead to permanent damage of the aortic orifice and to chronic aortitis. In the syphilitic forms the amount of change depends greatly on early diagnosis and proper treatment.

The **TREATMENT** is that of the etiological condition.

**Chronic Aortitis.**—(DILATATION OF THE AORTA).—This is a common condition, frequently overlooked. The diffuse dilatation is sometimes described under aneurism but deserves separate mention. It was first described by Hodgson in 1815 as “preternatural permanent enlargement of the cavity of an artery.” It is often associated with aortic insufficiency, a combination which the French term *maladie de Hodgson*.

**ETIOLOGY.**—It is much more common in males and the colored race shows a relatively high incidence. There are several special groups: (1) As a result of infection and acute aortitis a permanent dilatation remains. Two diseases are particularly concerned, rheumatic fever and syphilis. (2) As part of a general arterio-sclerosis in which the aorta is specially involved. The influence of syphilis and hard muscular work is important in this form. (3) In the aged it is common as part of an advanced arterial degeneration.

**PATHOLOGY.**—The extent of dilatation varies greatly and may involve only a portion of the arch, extend throughout the whole extent of the aorta or only to where the aorta passes through the diaphragm. The orifices and part of the vessels given off from the aorta may be involved in the dilatation. Thrombus formation in the aorta may occur. The aorta shows all grades of gross atheromatous change.

**SYMPTOMS.**—There are several groups: (1) Latent cases, especially in the aged. (2) In many cases the symptoms due to associated cardiac disease predominate, with the features of myocardial insufficiency or aortic insufficiency. (3) A group with features suggestive of angina pectoris, not surprising in view of the disease of the first part of the aorta. The pain may radiate down either arm or sometimes down both. The common complaints are of pain, dyspnoea, and cough.

**SIGNS.**—The neck may be full with distended veins and a collar of pulsation above the clavicles and sternum. Pulsation in the suprasternal notch is common. The manubrium may be lifted and pulsation in the upper two interspaces is often seen. The order of frequency is second right, second left, first right and first left interspace. This pulsation is usually diffuse and can rarely be felt distinctly. The aorta may be felt above the sternum or with the finger behind it. *Dulness* is very important, over the manubrium and adjoining interspaces. It is continuous with the heart dulness in most cases but not always. The width of the dulness in the first interspaces may be 8 to 14 cm. and the extent may vary from time to time. On *auscultation* the second sound often has an amphoric bell-like quality, which is diagnostic if present. The murmur of an associated aortic insufficiency may have the same quality. The blood pressure is low in the majority. Arterio-sclerosis is usually and aortic insufficiency (relative or permanent) often present. The *pressure signs* are practically the same as in aneurism, inequality of the pupils, laryn-

## ANEURISM

geal paralysis, tracheal tug, inequality of the radial pulses, and dysphagia.

**DIAGNOSIS.**—The main point is to know of the condition and look for it. The diagnosis from aneurism or displacement of the aorta is difficult in a few cases; the X-ray study will decide. The pain suggests angina pectoris but is rarely so severe, often lasts for a considerable time, and is not often caused by exertion; mild exertion often relieves the pain. Sweating is very rare.

**TREATMENT.**—A quiet even life with avoidance of strain, physical or mental, a limited diet, open bowels, and the treatment of symptoms are the main points. If syphilis is responsible, thorough treatment should be given but usually the damage is done and beyond repair. Vaso-dilators are useful for the pain.

## V. ANEURISM

**Definition.**—A tumor containing blood or blood clot in direct communication with the cavity of the heart, the surface of a valve, or the lumen of an artery.

**History.**—Galen knew external aneurism well, and in the second century A. D., Antyllus devised his operation of incising and emptying the sac inclosed between ligatures. Internal aneurism was recognized by Fernelius in the 16th century and Vesalius was very familiar with it. Ambroise Paré suggested the relation of aneurism to syphilis, which was insisted upon in the great monograph of Lancisi in 1728. Morgagni in 1761 described very fully the symptoms and morbid anatomy. The modern views date from the studies of Helmstedter and Köster, who showed that the primary change was in the media. The researches of Eppinger, Thoma, and Welch emphasized the importance of the changes in the media, particularly as due to syphilis.

**Classification.**—The following classification may be adopted:

I. **TRUE ANEURISM**, in which one or more of the coats of the vessel form the wall of the tumor: (a) *Dilatation-aneurism*—(1) Limited to a certain portion of the vessel, fusiform, cylindroid; (2) extending over a whole artery and its branches—*cirroid aneurism*. (b) *Circumscribed saccular aneurism*, which is the common form of aneurism of the aorta. (c) *Dissecting aneurism*, with splitting of the media, and occasionally with the formation of a new tube lined with intimal endothelium.

II. **FALSE ANEURISM**, following a wound or the rupture of an artery or of a true aneurism, causing a diffuse, or circumscribed, hæmatoma.

III. **ARTERIO-VEINOUS ANEURISM**, either with direct communication between an artery and vein, or with the intervention of a sac, *varicose aneurism*.

IV. **SPECIAL FORMS**, as the parasitic, erosion, traction, and mycotic.

**Etiology.**—**PREDISPOSING CAUSES.**—*Age.*—Nearly one-half of the deaths in England and Wales from aneurism in males occur between the ages of 30 and 45. In the young and in the very old the disease is rare, but it may occur at any age. Richy collected 41 cases (1923) of aneurism of the thoracic aorta, below the age of 19 years. Only two cases were syphilitic and nearly half were mycotic, usually with rheumatic fever. Congenital aneurism has been described.

*Sex.*—Males are attacked much more frequently than females—5 to 1.

*Race and Locality.*—Lucke and Rea in the collected statistics of 160,145 autopsies found 1,452 cases of aneurism (1 in 117). The proportion was higher in the United States than in Great Britain, Scandinavia, Germany or Austria. The disease is more common in Great Britain than on the Continent. Among about 19,000 post-mortems at Vienna there were 230 cases of aneurism, while among 18,678 at Guy's Hospital there were 325 cases. It is more common in the negroes of the southern states of America than among the whites. Of 345 admissions for aneurism to the Hopkins Hospital 132 were in colored and 213 in white patients—a ratio of 1 to 1.6, while the ratio of colored to white in the hospital at large was 1 to 5. In India aneurism is rare, though syphilis and arterial disease are common. Possibly, as Rogers suggests, the low blood pressure in the natives may have something to do with this comparative immunity.

*Occupation.*—Soldiers, sailors, draymen, iron and steel workers, and dock workers are particularly prone. This is due to the effect of strain on a vessel damaged by syphilis.

DETERMINING CAUSES.—These are three in number:

I. *The Acute Infections.*—In the specific fevers areas of degeneration are common in the aorta. Fortunately in most instances they are confined to the intima, but occasionally, as Thayer pointed out in typhoid fever, the changes may be in the media. The infection with which aneurism is especially connected is *syphilis*—a fact recognized in the eighteenth century by Lancisi and by Morgagni, and dwelt upon specially in 1876 by Francis H. Welch, of the British Army. All recent figures show a very high percentage of syphilis and it is rare not to find a positive Wassermann reaction in an aneurismal patient. Other infections play a very minor rôle. With rheumatic fever, pneumonia, and septicæmia, the mycotic aneurism may be associated.

II. The second factor is *strain*, particularly that associated with sudden and violent muscular effort. The media is the protecting coat of the artery, and during a violent effort, laceration or splitting of the intima may occur over a weak spot. If small this leads to a local bulging of the media and the gradual production of a sac, or the tear of the intima may heal completely, or a dissecting aneurism may form. In other instances a widespread mesoarteritis leads to gradual, diffuse distention of the artery. This type of aneurism, frequent in the aged, may follow ordinary chronic atheroma.

III. *Occasional Causes.*—(a) Embolism: The emboli may consist of vegetations or calcified fragments from the valves. This form, often multiple, is met with in infective endocarditis, in which the emboli probably pass to the vasa vasorum, causing mesoarteritis with weakening of the wall; but in the smaller vessels the aneurisms are caused by the direct lodgment of the emboli which infect and weaken the wall. (b) External Injury: A blow on the chest, a sudden fall, or the jar of an accident may cause a rupture of the intima over a weak spot in the aorta, with the production of a dissecting or sacculated aneurism. (c) External Erosion: A tuberculous focus may invade the wall of the aorta; or a bullet lodged near the wall of an artery may weaken it and be followed by aneurism. (d) In the horse there is a *parasitic aneurism* common in the mesenteric vessels, due to growth in them of the *Strongylus armatus*. (e) Thoma described a "traction" aneurism at the concavity of the arch at the point of insertion of the ductus Botalli.

**Morbid Anatomy.**—**NUMBER.**—Usually there is one aneurism, but three or four or even a dozen may be present. Multiple cup-shaped tumors in the aorta are always syphilitic. The mycotic are usually multiple, and in the peripheral vessels there may be a dozen or more.

**FORM.**—There are two great types—one in which the lumen of the vessel is dilated, and the other in which a limited section of the wall gives way with the formation of a sac. Typical cylindrical and spindle shaped aneurisms are seen in the aorta and in the vessels of the second and third dimensions. The sacculated form is the more common. They are either flat, saucer-shaped, or cup-shaped, or sometimes beyond a very narrow orifice is a cylindrical tumor of variable size, from a pin's head in the smaller vessels, as in the brain, to a huge sac which may fill one-half of the chest.

**VESSELS AFFECTED.**—Of a series of 551 cases studied by Crisp, the thoracic aorta was involved in 175, abdominal aorta in 59, femoral-iliac in 66, popliteal in 137, innominate in 20, carotids in 25, subclavians in 23, axillary in 18. The other smaller vessels are rarely attacked. Of late years aneurism of the external vessels appears to be much less frequent.

## I. ANEURISM OF THE AORTA

**A. Aneurism of the Thoracic Aorta.**—For purposes of discussion this part of the vessel may be divided into the sinuses of Valsalva, ascending, transverse, and descending portions.

(a) **ANEURISM OF THE SINUSES OF VALSALVA**, a common and important variety, is most frequent in young syphilitic subjects. There may be pouching of one or all three sinuses; the aortic ring is apt to be involved with aortic insufficiency. The special features are: (1) It is often *latent*, causing sudden death by perforation into the pericardium. (2) It is a medico-legal aneurism met with in coroner's cases. (3) Angina pectoris is not uncommon and may be the only symptom. (4) Aortic insufficiency is often associated with it. (5) In a majority of all cases syphilitic mesaortitis is present.

(b) **ANEURISM OF THE ASCENDING ARCH.**—Along the convex border aneurism frequently arises and may grow to a large size, either passing out into the right pleura or forward, pointing at the second or third interspace, eroding the ribs and sternum, and producing an external tumor. In this situation the sac may compress the superior vena cava, causing engorgement of the vessels of the head and arm; sometimes it compresses only the subclavian vein, and causes enlargement and edema of the right arm. Perforation may take place into the superior vena cava, of which Pepper and Griffith collected 29 cases. In rare instances, when the aneurism springs from the concave side of the vessel, the tumor may appear to the left of the sternum. Large aneurisms in this situation may cause much dislocation of the heart, pushing it down and to the left, and sometimes compressing the inferior vena cava, and causing swelling of the feet and ascites. The right recurrent laryngeal nerve is often compressed. The innominate artery is rarely involved. Death commonly follows from rupture into the pericardium, pleura, or superior vena cava; less commonly from rupture externally, sometimes from syncope.

(c) **ANEURISM OF THE TRANSVERSE ARCH.**—The direction of growth is most commonly backward, but the sac may grow forward, erode the sternum,

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and form a large tumor. The sac presents in the middle line and to the right of the sternum much more often than to the left, which occurred in only 4 of 35 aneurisms in this situation (O. A. Browne). Even when small and producing no external tumor it may cause marked pressure signs, involving the trachea and œsophagus, and giving rise to cough, often paroxysmal, and dysphagia. The left recurrent laryngeal is often involved in its course round the arch. A small aneurism from the lower or posterior wall of the arch may compress a bronchus, inducing bronchorrhœa, bronchiectasis, and sup-puration in the lung—a process which not infrequently causes death in aneurism, sometimes termed “aneurismal phthisis.” Occasionally enormous aneurisms arise in this situation, and grow into both pleuræ, extending between the manubrium and the vertebræ; they may persist for years. The sac may be evident at the sternal notch. The innominate artery, less commonly the left carotid and subclavian, may be involved, and the radial or carotid pulse absent or retarded. Sometimes the thoracic duct is compressed.

The ascending and transverse portions of the arch are not infrequently involved together, usually without the branches; the tumor grows upward, or upward and to the right.

(d) ANEURISM OF THE DESCENDING PORTION OF THE ARCH.—This may be the traction aneurism of Thoma. The sac projects to the left and backward, and often erodes the vertebræ from the third to the sixth dorsal, causing great pain and sometimes compression of the spinal cord. Dysphagia is common. Pressure on a bronchus may induce bronchiectasis, with retention of secretions, and fever. A tumor may appear externally in the region of the scapula, and attain an enormous size. Death may occur from rupture into the pleura, or the sac may grow into the lung and cause hæmoptysis.

(e) ANEURISM OF THE DESCENDING THORACIC AORTA.—This is the least common situation of aortic aneurism. The larger number occur close to the diaphragm, the sac lying upon or to the left of the bodies of the lower dorsal vertebræ, which are often eroded. It may be latent, in 3 of 14 cases (Osler), and is often overlooked; pulmonary and pleural symptoms are common. Pain in the back is severe; dysphagia is not infrequent. Slow leaking may cause symptoms suggestive of an acute abdominal condition. The sac may reach an enormous size and form a subcutaneous tumor in the left back.

**Symptoms.**—Broadbent made the useful division of aneurisms of *symptoms* and aneurisms of *physical signs*; the former is more commonly seen when the transverse arch is involved, the latter when the ascending portion. There may be no symptoms. A man may present a tumor which has eroded the chest wall without pain or any discomfort but this is rare.

An important feature in thoracic aneurism is *pain*, which is particularly marked in deep seated tumors. It is usually paroxysmal, sharp, and lancinating, often very severe when the tumor is eroding the vertebræ, or perforating the chest wall. In the latter case after perforation the pain may cease. Anginal attacks are not uncommon, particularly in aneurisms at the root of the aorta. Frequently the pain radiates down the left arm or up the neck, sometimes along the upper intercostal nerves. Superficial tenderness may be present over the heart or over the left sterno-mastoid muscle. *Cough* results either from the direct pressure on the trachea or is associated with bronchitis. The expectoration in these instances is abundant, thin, and watery; subse-

quently it becomes thick and turbid. *Paroxysmal cough* of a peculiar brazen, ringing character is a characteristic feature in some cases, particularly when there is pressure on the recurrent laryngeal nerves; the cough may have a peculiar wheezy quality—the “goose cough.”

*Dyspnea*, which is common in aneurism of the transverse portion, is not necessarily associated with pressure on the recurrent laryngeal nerves, but may be due directly to compression of the trachea or a bronchus. It may occur with marked stridor. Loss of voice and hoarseness are consequences of pressure on the recurrent laryngeal, usually the left, inducing paralysis or spasm of the muscles of the left vocal cord. Paralysis of an abductor on one side may be present without any symptoms. It is more particularly, as Semon states, when paralytic contractures supervene that attention is called to laryngeal symptoms.

*Dysphagia* is comparatively rare and may be due to spasm or direct compression. A tube or sound should never be passed in these cases, as the oesophagus may be almost eroded and perforation of the sac has taken place.

*Heart Symptoms.*—Pain is often anginal in character, and is most common when the root of the aorta is involved. The heart is hypertrophied in less than one-half of the cases. The aortic valves are sometimes incompetent, either from disease of the segments or stretching of the aortic ring.

**Physical Signs.**—**INSPECTION.**—A good light is essential; cases are often overlooked owing to a hasty inspection. The face is often suffused, the conjunctivæ injected, and veins of the chest and of one arm engorged. The pupils may be unequal. In many instances inspection is negative. On either side of the sternum there may be abnormal pulsation which should be looked for carefully. Three sorts of pulsation may be seen in the chest: (1) A general shock, such as is seen in violent throbbing of the heart or of an aneurism. In anæmia, in neurasthenia, and in great hypertrophy this widespread shock may suggest aneurism. (2) A diffuse localized impulse which may be caused by a deep-seated aneurism but which is met with also in tumors, in pulsating pleurisy, and in a few cases without evident cause. (3) The punctate, heaving true aneurismal impulse which when of any extent is visibly expansile. It is seen most frequently above the level of the third rib to the right of the sternum, in the second left interspace, over the manubrium, and behind in the left interscapular region. When the innominate is involved the throbbing may be seen at the right sterno-clavicular joint and above it. An external tumor is present in many cases, projecting either through the upper part of the sternum or to the right, sometimes involving the sternum and costal cartilages on both sides, forming a swelling the size of a cocoanut or even larger. The skin is thin, often blood stained, or it may have ruptured, exposing the laminæ of the sac. The apex beat may be displaced, particularly when the sac is large. It is more commonly due to dislocation than to enlargement of the heart.

**PALPATION.**—The area and degree of pulsation are sometimes best determined by palpation. When the aneurism is deep seated and not apparent externally, the bimanual method should be used, one hand upon the spine and the other on the sternum. There may be only a diffuse impulse. When the sac has perforated the chest wall the impulse is, as a rule, forcible, slow, heaving, and expansile. It may be more forcible than the apex beat. The resist-

ance may be great if there are thick laminae beneath the skin; more rarely the sac is soft and fluctuating. The hand upon the sac, or on the region in which it is in contact with the chest wall, may feel a diastolic *shock*, often of great intensity, which is a valuable sign of aneurism. A systolic thrill is sometimes present. The pulsation is sometimes felt in the suprasternal notch.

**PERCUSSION.**—The small and deep seated aneurisms are in this respect negative. In the larger tumors, as soon as the sac reaches the chest wall, there is an area of abnormal dullness, the position of which depends upon the part of the aorta affected. Aneurisms of the ascending arch grow forward and to the right, producing dullness on one side of the manubrium; those from the transverse arch produce dullness in the middle line, extending toward the left of the sternum, while aneurisms of the descending portion most commonly produce dullness in the left interscapular and scapular regions. The percussion note is flat and there is a feeling of increased resistance.

**AUSCULTATION.**—Adventitious sounds are not always heard. Even in a large sac there may be no murmur. Much depends upon the thickness of the laminae of fibrin. An important sign, particularly if heard over a dull region, is a ringing, accentuated second sound. A systolic murmur may be present; sometimes a double murmur, in which case the diastolic is usually due to aortic insufficiency. A systolic murmur alone is of little moment in the diagnosis of aneurism. A continuous humming top murmur with systolic intensification is heard when the aneurism communicates with the vena cava or the pulmonary artery.

Among OTHER PHYSICAL SIGNS of importance are retardation of the pulse in the arteries beyond the aneurism, or in those involved in the sac. There may be a marked difference between the right and left radial, both in volume and time. The blood pressure on the two sides may be unequal. A sign of large thoracic aneurism is obliteration of the pulse in the abdominal aorta and its branches. Attention was called to this in a patient who was stated to have aortic insufficiency. There was a well-marked diastolic murmur, but in the femorals and abdominal aorta no trace of pulsation could be found. A careful examination of the patient's back showed a very large area of pulsation in the left scapular region. The sac probably was large enough to act as a reservoir annihilating the pulse wave and converting the intermittent into a continuous stream.

A condition suggestive of pneumo-thorax may be caused by compression of one bronchus by the sac (Newton Pitt). Air passes beyond the obstruction, but has difficulty in getting out, so that the lung is gradually distended, causing enlargement of the side with a hyperresonant note and absence of breath sounds. The X-ray picture may alone decide the diagnosis.

The *tracheal tugging*, a valuable sign in deep-seated aneurisms, was described by Surgeon-Major Oliver. The patient should be in the erect position with the mouth closed and the chin low. The cricoid cartilage is grasped between the finger and thumb or the ends of two fingers are hooked under it and steady pressure made upward. A pull or tug on the trachea is felt. Sometimes it is visible. This is a sign of great value in the diagnosis of deep-seated aneurisms, though it may occasionally be felt in tumors, in the extreme pulsation of aortic insufficiency and in dilatation of the aorta. The trachea may be pushed to one side.

Occasionally a systolic murmur may be heard in the trachea, as pointed out by David Drummond; or even at the patient's mouth, when opened. This is either the sound conveyed from the sac, or is produced by the air as it is driven out of the trachea during systole. Altered physical signs in one lung are common effects of pressure.

*Hæmorrhage* may come from (a) soft granulations in the trachea at the point of compression, in which case the sputum is blood tinged, but large quantities of blood are not lost; (b) from rupture of the sac into the trachea or a bronchus; (c) from perforation into the lung or erosion of the lung tissue. The bleeding may be profuse, rapidly proving fatal, and is a common cause of death. It may persist for weeks or months, in which case it is simply hæmorrhagic weeping through the sac, which is exposed in the trachea. In some instances, even after a profuse hæmorrhage, the patient recovers and may live for years. A man with well-marked thoracic aneurism, who had several brisk hæmorrhages, died four years after, having in the meantime enjoyed average health. Death from hæmorrhage is relatively more common in aneurism of the third portion of the arch and of the descending aorta.

Among other signs, *venous compression* may involve one subclavian or the superior vena cava. A curious phenomenon is the clubbing of the fingers of one hand, of which two examples were without any special distention or signs of venous engorgement. Tumors of the arch may involve the pulmonary artery, producing compression, or in some instances adhesion of the pulmonary segments and insufficiency of the valve; or the sac may rupture into the artery, producing instantaneous death.

*Pupil Signs.*—These may be due to pressure on the *sympathetic*, which may cause dilatation of one pupil from irritation, contraction when the nerve is paralyzed. Flushing of the side of the face and ear, increased temperature, and sweating may be present. As Ainley Walker and Wall have shown, the anisocoria is frequently due to vascular conditions—with low blood pressure in one carotid the pupil on that side is dilated, with high pressure contracted, and in 26 cases of aneurism they found a relation between the state of the pupil and the arteries on the same side. In some cases the anisocoria is a syphilitic manifestation.

An X-ray study should be made in all doubtful cases. The fluoroscope gives an accurate picture of the situation, the size, and the relation to the heart. Even a small sac may be seen. The diagnosis may rest upon it in cases in which scarcely a physical sign is present. Sailer and Pfahler have shown that tortuosity of the aorta, due to arterio-sclerosis, may exist, suggesting aneurism, particularly with the fluoroscope.

The clinical picture of aneurism of the aorta is extremely varied. Many cases present characteristic symptoms and no physical signs, while others have well-marked physical signs and few or no symptoms.

**Diagnosis.**—Aneurism of the aorta may be confounded with: (a) The violent throbbing impulse of the arch in aortic insufficiency.

(b) *Simple Dynamic Pulsation.*—This is common in the abdominal aorta, but rare in the arch. A case under the care of Murray and Bramwell presented, without pain or pressure signs, pulsation and dulness over the aorta. The condition gradually disappeared and was thought to be neurotic.

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(c) *Dilatation of the arch* has many of the features of aneurism. The X-ray examination may be required to decide the diagnosis.

(d) In *curvature of the spine* there may be great displacement of the aorta, so that it pulsates forcibly to the right of the sternum.

(e) *Solid Tumors*.—When the tumor projects externally and pulsates the difficulty may be considerable. In tumor the heaving, *expansile* pulsation is absent, and there is not that sense of force and power which is so striking in the throbbing of a perforating aneurism. A diastolic shock is not felt. Auscultatory signs are less definite, as large aneurisms may occur without murmurs and murmurs may be heard over tumors. The greatest difficulty is in the deep seated thoracic tumors, and here the diagnosis may be impossible. The physical signs may be indefinite. The ringing aortic second sound is rarely, if ever, heard over tumor. Tracheal tugging is a valuable sign. Pressure phenomena are less common in tumor. The general appearance of the patient in aneurism is much better than in tumor, in which there may be cachexia and enlargement of the glands in the axilla or in the neck. The result of the Wassermann reaction is of aid. Occasionally cancer of the oesophagus simulates aneurism, producing pressure on the left bronchus.

(f) *Pulsating Pleurisy*.—In *empyema necessitatis*, if the projecting tumor is in the neighborhood of the heart and pulsates, the condition may be mistaken for aneurism. The absence of a heaving, firm impulse and of a diastolic shock would, with the history and the existence of pleural effusion, determine the diagnosis. If necessary, puncture may be made with a fine needle. In a majority of the cases of pulsating pleurisy the throbbing is diffuse and widespread, moving the whole side. The X-ray study is of value.

**Prognosis.**—The outlook is always grave. Life may be prolonged for some years, but the patients are in constant jeopardy. Spontaneous cure is not very infrequent in the small sacculated tumors. The cavity becomes filled with laminae of firm fibrin, which become more and more dense and hard, the sac shrinks considerably, and finally lime salts are deposited in the old fibrin. The laminae of fibrin may be on a level with the lumen of the vessel, causing complete obliteration of the sac. The cases which rupture externally, as a rule, run a rapid course, although to this there are exceptions; the sac may contract, become firm and hard, and the patient may live for five, or even ten years or longer. The cases which last longest are those in which a saccular aneurism projects from the ascending arch. The aneurism may be enormous, occupying a large area of the chest, and yet life be prolonged for years. One of the most remarkable instances is the case of dissecting aneurism reported by Graham. The patient was invalided after the Crimean War with aneurism of the aorta, and for years was under the observation of J. H. Richardson, of Toronto, under whose care he died in 1885. The autopsy showed a healed aneurism of the arch, with a dissecting aneurism extending the whole length of the aorta, which formed a double tube.

**Treatment.**—In a large proportion of cases this can only be *palliative* but in every instance measures should be taken which promote clotting and consolidation in the sac. In any series of cured aneurisms a majority of the patients have not been known to be subjects of the disease, but the obliterated sac has been found accidentally at the post mortem.

The most satisfactory plan in early cases, when it can be carried out

thoroughly, is the modified Valsalva method advised by Tufnell, of Dublin, the essentials of which are rest and a restricted diet. The rest should, as far as possible, be absolute. The reduction of the daily number of heart beats, when a patient is recumbent and without exertion, amounts to many thousands, and is one of the principal advantages of this plan. Mental quiet should also be secured. The diet advised by Tufnell is extremely rigid—a total of 10 ounces of solid food and 8 ounces of fluid a day. It is not possible to keep to such amounts for any long period but the intake should be as small as possible. The bowels should be kept freely open and straining at stool avoided. This treatment should be pursued for several months, but usually it is impossible to carry it out for more than a few weeks at a time. It is a form of treatment adapted only to the saccular form, and in large sacs communicating with the aorta by a comparatively small orifice the chances of consolidation are fairly good. The patient with symptoms should be at rest on a low diet. At all times he should live a quiet life, avoiding over-exertion, fatigue and strain. The bowels should be kept regular, and constipation and straining carefully avoided. Of medicines, iodide of potassium is of great value. It may be given in doses of from 5 to 20 grains (0.3 to 1.3 gm.) three times a day. Larger doses are not necessary. The most striking effect of the iodide is the relief of pain. Although the damage is done, yet it is well to give specific treatment. Neo-arsphenamine should be given very cautiously at first and the effect carefully watched. There is risk with its use and the initial dose should be not more than 0.1 gram. A thorough course of mercury should be given and repeated as often as is indicated.

In cases in which the tumor is large, or in which there seems little prospect of consolidation, it is perhaps better to advise a man to go on quietly with his occupation, avoiding excitement and worry. Our profession has offered many examples of good work by men with aneurism of the aorta, who wisely preferred, as did the late Hilton Fagge, to die in harness.

OTHER CONDITIONS.—Pressure on veins causing engorgement, particularly of the head and arms, is sometimes promptly relieved by free *venesection*, and, at any time, if attacks of dyspnoea with lividity supervene, bleeding may be resorted to with great benefit. In the final stages morphia is, as a rule, necessary. Dyspnoea, if associated with cyanosis, is best relieved by bleeding. Chloroform inhalations may be necessary. The question of tracheotomy sometimes comes up in the cases of urgent dyspnoea. If it can be shown by laryngoscopic examination that it is due to bilateral abductor paralysis the trachea may be opened, but this is extremely rare, and urgent dyspnoea is usually caused by pressure about the bifurcation. When the sac appears externally and grows large, an ice bag or a belladonna plaster may be applied to allay the pain but wiring and electrolysis are most useful for this. In some instances an elastic support may be used with advantage, and a physician with an enormous external aneurism in the right mammary region obtained great relief by an elastic support, passing over the shoulder and under the arm of the opposite side.

The nitrites may be given if the blood pressure is high, but rest and diet, restriction of the fluids, and free purgation are usually more effectual than drugs in reducing blood pressure.

**SURGICAL MEASURES.**—Consolidation may be promoted in the sac by the combination of wiring and electrolysis. Moore, in 1864, first wired a sac, putting in 78 feet of fine wire. Death occurred on the fifth day. Corradi proposed the combined method of wiring with electrolysis, which was first used by Burresi in 1879. H. A. Hare has done the operation 32 times without any accident. He emphasizes the importance of employing a gold platinum wire without too much spring (silver is not suitable), of using the positive pole in the aneurism and of not giving too strong a current (5 milliamperes at the beginning, gradually increased to 50, and then decreased to 5 again, the current being passed for about 50 minutes). In nearly all of Hare's patients there was marked benefit, the duration of which was variable. One patient lived for nine years. The decrease in the size of the aneurism is often marked but the relief of pain is the most striking feature. The most favorable cases are those in which the aneurism is sacculated, which can usually be determined by the X-rays.

**B. Aneurism of the Abdominal Aorta.**—Of 233 cases collected by Nixon, 207 were in males, 26 in females; 121 were between the ages of twenty-five and forty-five. Nixon reports a case in a syphilitic girl of twenty. There were 16 cases among 16,000 admissions at the Johns Hopkins Hospital.

**Pathology.**—The sac is most common just below the diaphragm in the neighborhood of the celiac axis. The tumor may be fusiform or sacculated, and it is sometimes multiple. Projecting backward, it erodes the vertebrae and may cause numbness and tingling in the legs and finally paraplegia, or it may pass into the thorax and burst into the pleura. More commonly the sac is on the anterior wall and projects forward as a definite tumor in the middle line or a little to the left. The tumor may project in the epigastric region (most common), in the left hypochondrium, in the left flank, or in the lumbar region. When high up beneath the pillar of the diaphragm it may attain considerable size without being very apparent on palpation. If it ruptures into the retro-peritoneal tissues a tumor in the flank may be formed gradually, which enlarges with very little pulsation. It may be mistaken for a rapidly growing sarcoma or for abscess, and an operation may be performed.

The *symptoms* are chiefly pain, very often of a neuralgic nature, passing round to the sides or localized in the back, and more persistent and intense than in any other variety of aneurism. Gastric symptoms, particularly vomiting, may be early and deceptive features.

**Diagnosis and Physical Signs.**—Inspection may show marked pulsation in the epigastric region, sometimes a definite tumor. A thrill is not uncommon. The pulsation is forcible, expansile, and sometimes double when the sac is large and in contact with the pericardium. On palpation a *definite tumor can be felt*. Though usually fixed, the aneurism may be freely movable. If large, there is some degree of dullness on percussion, which usually merges with that of the left lobe of the liver. On auscultation, a systolic murmur is, as a rule, audible, and is sometimes best heard at the back. A diastolic murmur is occasionally present, usually very soft in quality. No pulsation, however forcible, no thrill, however intense, no murmur, however loud, justifies the diagnosis of abdominal aneurism unless there is a *definite tumor which can be grasped and which has an expansile pulsation*. Attention to this rule

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will save many errors. Retardation of the pulse in the femoral artery is common. The *throbbing abdominal aorta* was well described by Morgagni and Laennec, and called by Allan Burns the "preternatural pulsation in the epigastrium." It is met with (a) in nervous women often associated with enteroptosis and pain, and sometimes, as Morgagni pointed out, with vomiting of blood. (b) In anæmia particularly after severe hæmorrhage, in which the throbbing may shake the patient and the bed. (c) In aortic insufficiency. (d) In sclerosis of the abdominal aorta. A common mistake is to regard this throbbing aorta as aneurism. The vessel may appear dilated and even may be grasped in the hand. Very frequently a tumor of the pylorus, of the pancreas, or of the left lobe of the liver is lifted with each impulse of the aorta and may be confounded with aneurism. The absence of the forcible expansile impulse and the examination in the knee elbow position, in which the tumor, as a rule, falls forward, and the pulsation is not then communicated, suffice for differentiation.

**Prognosis.**—The outlook is bad but a few cases heal spontaneously. Death may result from (a) complete obliteration of the lumen by clots; (b) compression paraplegia; (c) rupture (which occurred in 152 of the 233 cases in Nixon's series) either into the pleura, retro-peritoneal tissues, peritoneum, or the intestines, most commonly into the duodenum; (d) embolism of the superior mesenteric artery, producing intestinal infarction.

The **treatment** is the same as in thoracic aneurism. With an aneurism low down pressure was successfully applied by Murray, of Newcastle. It must be kept up for many hours under anaesthesia. The plan is not without risk, as patients have died from bruising and injury of the sac. Nine cases in our series were treated surgically. In two the wiring and electrolysis were followed by great improvement; one man lived for three years.

**C. Dissecting Aneurism.**—The majority of aneurisms of the aorta begin with a split or crack of the intima over a spot of syphilitic mesaortitis. Once this split has started the aorta may rupture in all its coats, or an aneurism may form at the site, or the fracture of the intima, though large and often circumferential, may heal; or the blood may extend between the coats, separating them for many inches, or in the entire extent, forming a dissecting aneurism; and, lastly, such a dissecting aneurism may heal perfectly.

RUPTURE OF THE AORTA is not very infrequent. Usually there is agonizing pain with features of shock, and death may take place instantly; but in fully half of the cases there are two very characteristic stages, the first corresponding to the rupture of the inner coats, the second eight to ten hours, or as long as fifteen days later, to fatal rupture of the external layer.

*Dissecting aneurism* is uncommon. There were only two cases in 16 years at the Hopkins Hospital, where aneurism is exceptionally frequent. The primary split is most frequently in the arch, not far above the valves, and is in the form of a transverse, or vertical, clean cut incision, as if made with a razor. The extent of the separation of the coats is variable. If the adventitia is reached, rupture is certain to take place, as only the structures of the middle coat can resist for any time the pressure of the blood. The blood may pass for three or four or more inches, separating the media, and then burst internally or externally. In other cases the dissection reaches from the ascending arch to the bifurcation of the aorta, even passing down

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the iliac and femorals into the vessels of the leg. The splitting of the coats may reach to all the subdivisions of the aorta. The symptoms are those spoken of under rupture; but a remarkable condition may follow, leading to:

**HEALED DISSECTING ANEURISM.**—The earlier observers of this remarkable condition regarded it as an anatomical anomaly of a double aorta. Adami collected 39 cases, in a majority of which there was no advanced disease of the aorta itself. The outer tube formed by the dissecting aneurism may extend the entire length of the aorta, occupying the full extent of the circumference. The most extraordinary feature is that the outer tube may present a perfectly smooth and natural appearance, and be lined with a new intima. The condition may last for many years.

### II. ANEURISM OF THE BRANCHES OF THE ABDOMINAL AORTA

The **coeliac axis** is not infrequently involved in aneurism of the first portion of the abdominal aorta. Of its branches, the **splenic artery** is occasionally the seat of aneurism. This rarely causes a tumor large enough to be felt; sometimes, however, the tumor is of large size. In a man, aged thirty, who had an illness of several months' duration, the severe epigastric pain and vomiting led to a diagnosis of gastric ulcer. There was a deep seated tumor in the left hypochondriac region, the dulness of which merged with that of the spleen. There was no pulsation, but it was thought on one occasion that a murmur was heard. The chief symptoms were vomiting, severe epigastric pain, occasional hæmatemesis, and finally severe hæmorrhage from the bowels. An aneurism of the splenic artery the size of a coconut was situated between the stomach above and the transverse colon below. The sac contained densely laminated fibrin. It had perforated the colon. Of 39 instances of aneurism on the branches of the abdominal aorta collected by Lebert, 10 were of the splenic artery.

Of aneurism of the **hepatic artery** Friedenwald and Tannenbaum collected 65 cases (1922) of which 45 were extra-hepatic. Rupture took place in 45 cases, in 33 into the abdominal cavity and in 21 into the bile passages. Infections other than syphilis play the main part in etiology. Pain, gastrointestinal hæmorrhages, jaundice and fever are common features, so that it is usually mistaken for gall-bladder disease or ulcer. Ligation of the artery may be done but is rarely successful.

Aneurism of the **superior mesenteric artery** is not very uncommon. The diagnosis is scarcely possible from aneurism of the aorta. Plugging of the branches or of the main stem may cause infarction of the bowel.

**Renal Artery.**—Henry Morris collected 21 instances of aneurism, 12 of which arose from injury. Many of them were false. Pulsation and a *bruit* are not always present. Four cases were operated upon; three recovered. In a case of Keen's the tumor and the kidney were removed together.

**Pulmonary Artery.**—Primary aneurism of the trunk is very rare. The forms are: (a) Of the trunk and main branches Henschen to 1906 collected 42 cases; and Possett (1909) added 9. Most of the patients were in the third and fourth decades, and syphilis is the important factor. Warthin demonstrated spirochaetes in athero-sclerosis with aneurism. (b) Acute embolic aneurism, which may be multiple in connection with septic thrombi in the

veins or endocarditis of the right side of the heart. (c) The small aneurisms in the walls of pulmonary cavities, already considered.

### III. MYCOTIC OR BACTERIAL ANEURISMS

**Etiology.**—As the name implies these are due to bacterial infection and have two modes of origin. (1) Intravascular. In the majority the infection comes from bacterial endocarditis, in 187 of a series of 217 cases collected by Stengel and Wolferth (1923). Other sources were infections in the lungs and bones. The most frequent organisms are non-hamolytic streptococci. Emboli play an important part, being carried in the vasa vasorum, or lodging, especially at a bifurcation, and infecting the wall. Direct infection by bacteria in the blood stream is rare. (2) Extravascular. In this the infection may come from an abscess or an infected lymph-node. The adventitia is first affected. The ages in the series mentioned varied from 4 to 74 years; the majority were in the second, third and fourth decades.

**Pathology.**—In the series of 217 cases, in 49 cases more than one aneurism was found. The aorta was involved in 66 cases, especially in the arch; the intracranial arteries in 34; the pulmonary artery in 6 and its branches in 8 cases. The size is usually small, not larger than a walnut, and rupture occurs frequently. The intima may be quite normal to the edge of the aneurism.

**Symptoms.**—Sometimes the occurrence of embolism is noted first, followed very soon by an aneurism, or the aneurism may be discovered first. There is usually severe pain with local redness and swelling. Expansile pulsation may be present. The condition may be mistaken for phlebitis at first. Rupture of the aneurism is common.

**Treatment.**—If the aneurism occurs with endocarditis the outlook is hopeless and symptomatic treatment is indicated. With an infection which is mild or subsiding operation may be done on a superficial artery. In some cases the aneurism has disappeared. Stengel and Wolferth report a case of healing of a mycotic aneurism.

### IV. ARTERIO-VEINOUS ANEURISM

In this form, known to Galen, but first accurately described by the great William Hunter, there is abnormal communication between an artery and a vein. When a tumor lies between the two it is known as *varicose aneurism*; when there is a direct communication without tumor the vein is chiefly distended and the condition is known as *aneurismal varix*.

While it occurs in the aorta, it is much more common in the peripheral arteries as a result of stab or gunshot wounds.

An aneurism of the ascending portion of the aorta may open directly into the vena cava; 29 cases of this lesion were analyzed by Pepper and Griffith. Cyanosis, edema, and great distention of the veins of the upper part of the body are the most frequent symptoms, and develop, as a rule, with suddenness. A thrill is present in some cases. A continuous murmur with systolic intensification is of great diagnostic value. Thurnam (*Medico-Chirurgical Transactions*, 1840) gave the first accurate account of this murmur and of this characteristic type of cyanosis. There is only one condition with

which it could be confounded, viz., the remarkable cyanosis of the upper part of the body which follows crushing accidents to the thorax. Perforation between the aorta and pulmonary artery causes very much the same symptoms. In a few cases an aneurism of the abdominal aorta perforates the inferior vena cava—œdema and cyanosis of the legs and lower half of the body, and the distinctive thrill and murmur are present.

In the *arterio-venous* aneurisms which follow stab and bullet wounds of the peripheral arteries the features are most characteristic. First, the veins enlarge as the arterial blood flows under high pressure into them. The affected limb may be greatly swollen and in a young person may lengthen, and the growth of hair is increased. Secondly, a strong *thrill* is felt, of maximum intensity at the site of the aneurism, but sometimes to be felt at the most distant parts of a limb. Thirdly, the characteristic continuous *murmur* with systolic intensification is heard. In the external arteries the condition may persist for years before disability is caused by enlargement of the veins and swelling of the limb. Surgical treatment by a skilled operator is indicated.

#### *Periarteritis Nodosa*

An inflammatory lesion of the smaller arteries, beginning in the outer coats, with hyaline degeneration of the media, and formation of secondary aneurisms with thrombosis and rupture. The nodular syphilitic arteritis should not be included in this group. Described first by Kussmaul and Maier, it has been made the subject of special study by Dickson, Longcope, Lamb, and Klotz. Some 42 cases are on record.

The *etiology* is uncertain. Most of the cases are in males of middle age and syphilis has been noted in a few cases. The disease appears to be a subacute infection with forms of staphylococci and streptococci (Klotz). The smaller arteries are involved, the branches of the cœliac axis, the mesenteric, the renal, hepatic, coronary, and more rarely those of the skin, lungs and brain. The nodular tumors vary in numbers from a dozen or more to many hundreds and are usually visible to the naked eye. They differ in structure from the other forms of nodular arteritis, the syphilitic and mycotic. The subcutaneous nodules present in eight cases led to the diagnosis in two.

The disease runs a course with mild fever, weakness, anæmia, muscular and joint pains, epigastric pain, vomiting, diarrhœa and purpura. Tonsillitis has not infrequently preceded the attack. The duration is from a few weeks to three or four months. Recovery has occurred.

## SECTION XI

# DISEASES OF THE GLANDS OF INTERNAL SECRETION

**Introduction.**—Disturbances in the endocrine glands may be due to hyper-, hypo- or dysfunction. The results may be shown in various ways: (1) the features caused by disturbance in the gland specially involved, (2) secondary disturbances in other endocrine glands, as they are all bound together, causing a pluriglandular syndrome, and (3) involvement of the sympathetic nervous system and, through this, widespread influence on many organs. There seem to be special relations between certain glands, which may take the form of inhibition or of stimulation. It is evident that the pluriglandular syndromes may present very complex problems.

The thyroid, pituitary and adrenal glands have a close relation with the sympathetic system and the sex glands, and are opposed to the pancreas in metabolism. As regards development, they are opposed to the thymus and pineal glands. The endocrine glands influence growth, metabolism and many bodily processes, especially with the sympathetic nervous system. The thyroid is stimulated by the sympathetic system proper, and it lowers the threshold of sympathetic stimulation. In early life it may be that the thymus acts as a check; if it persists, certain changes result. The thyroid and pituitary particularly influence growth until the sex glands become active. In later life the acceleration glands, such as the thyroid, tend to have less influence. Throughout there may be disturbing factors of which psychological, metabolic, and toxic, especially bacterial, are the most important. The factors originating in the psychological life act through the sympathetic system, the metabolic in various ways, often through nutrition, and the toxic by influencing the glands and their secretions. In all a vicious circle may be established.

## I. DISEASES OF THE SUPRARENAL BODIES AND CHROMAFFIN SYSTEM

**Introduction.**—Of the two parts of the suprarenal bodies, (1) the *medullary* belongs to what is known as the chromaffin system, which includes a similar tissue scattered in the sympathetic ganglia and the carotid glands, and (2) the cortex with an epithelial origin and belonging to the interrenal system. The chromaffin bodies produce an internal secretion, *epinephrine*, supposed by some to maintain the blood pressure and the sympathetic tonus. Epinephrine is regarded as a stimulant to the sympathetic system, as having a vaso-constrictor effect on the blood-vessels, except the coronary arteries on which it has a dilator effect, and a dilator effect on the bronchi; it also causes an increase in the amount of sugar in the blood. In some way it influences the pigment

## DISEASES OF THE GLANDS OF INTERNAL SECRETION

metabolism of the skin and possibly the muscular vigor. Disturbance in function of the medullary portion of the suprarenal bodies is known only through the disease described by Addison. Beyond this all is debatable, and much visionary. The function of the cortical part of the gland is unknown, but that it bears some relation to the sexual organs is shown by the sex anomalies that develop with tumors of these parts and by the enlargement during pregnancy. Hyperplasia of the cortex or tumor formation may be associated with precocious sexual development and hypoplasia with infantilism. The interrenal system produces *cholin* which lowers blood-pressure.

*Glycosuria* is caused by the injection of epinephrine, and in animals a form of arterio-sclerosis, probably due to the high blood pressure. Many theoretical conceptions have been entertained of the relation between a defect of the adrenal secretion and asthenic affections, and it is suggested that adrenal insufficiency plays an important rôle in acute infections, in tuberculosis, and many wasting diseases, with which it is interesting to note that increased pigmentation may be associated.

### I. ADDISON'S DISEASE

**Definition.**—A disease characterized by muscular and vascular asthenia, gastro-intestinal disturbance, and pigmentation of the skin; due either to tuberculosis or atrophy of the medulla of the adrenals, or to degenerative changes in the chromaffin system generally.

The recognition of the disease is due to Addison of Guy's Hospital, whose monograph on "The Constitutional and Local Effects of Disease of the Suprarenal Capsules" was published in 1855.

**Etiology.**—The disease is rare. Only 17 cases were seen in 21 years in the United States (Osler). In large clinics a year or more may pass without a case. Males are more frequently attacked than females. In Greenhow's analysis of 183 cases, 119 were males and 64 females. The majority of cases occur between the twentieth and fortieth years. A congenital case has been described, in which the child lived for eight weeks and post mortem the adrenals were found to be large and cystic. In a few cases a blow on the abdomen or back has preceded the onset. A certain number of cases have been associated with Pott's disease. An increase in the disease in France was reported during the Great War.

**Morbid Anatomy.**—There is rarely emaciation or anæmia. Rolleston thus summarizes the condition of the suprarenal bodies in Addison's disease:

"1. The fibro-caseous lesion due to tuberculosis—far the commonest condition found. 2. Simple atrophy. 3. Chronic interstitial inflammation leading to atrophy. 4. Malignant disease invading the capsules, including Addison's case of malignant nodule compressing the suprarenal vein. 5. Blood extravasated into the suprarenal bodies. 6. No lesion of the suprarenal bodies themselves, but pressure or inflammation involving the semilunar ganglia.

"The first is the only common cause of Addison's disease. The others, with the exception of simple atrophy, may be considered as very rare."

The nerve-cells of the semilunar ganglia have been found degenerated and deeply pigmented, and the nerves sclerotic. The ganglia are not uncom-

## SUPRARENAL BODIES AND CHROMAFFIN SYSTEM

monly entangled in the cicatricial tissue about the adrenals. The chromaffin cells in the sympathetic ganglia and in the abdominal plexuses generally disappear. The cases of extensive destruction of the glands without Addison's disease are explained by a persistence of the chromaffin structures elsewhere, while extensive involvement of the extra-capsular chromaffin system may be sufficient to cause the symptoms, the adrenals themselves being intact.

Few changes of importance are found in other organs. The spleen is occasionally enlarged; the thymus may be persistent and the lymph-nodes and tonsils enlarged. The thyroid gland may show changes. The other organs show only the alterations associated with a protracted illness.

**Symptoms.**—In the words of Addison, the characteristic symptoms are "anæmia, general languor or debility, remarkable feebleness of the heart's action, irritability of the stomach, and a peculiar change of color in the skin." The onset is, as a rule, insidious. The feelings of weakness usually precede the pigmentation. In other instances the gastro-intestinal symptoms, the weakness, and the pigmentation come on together. There are a few cases in which the whole process is acute, following a shock or some special depression. There are several important features:

(a) **PIGMENTATION OF THE SKIN.**—This, as a rule, first attracts the attention of the patient's friends. The grade of coloration ranges from a light yellow to a deep brown, or even black. In typical cases it is diffuse, but always deeper on the exposed parts and in the regions where the normal pigmentation is more intense, as the areolæ of the nipples and about the genitals; also wherever the skin is compressed or irritated. At first it may be confined to the face and hands. Occasionally it is absent. Patches showing atrophy of pigment, leucoderma, may occur. The pigmentation occurs on the mucous membranes of the mouth, conjunctivæ, and vagina but it is not distinctive and is common in the negro. A patchy pigmentation of the serous membranes has often been found. Over the diffusely pigmented skin there may be little mole-like spots of deeper pigmentation, and upon the trunk, particularly on the lower abdomen, it may be "ribbed" like the sand on the seashore.

(b) **GASTRO-INTESTINAL SYMPTOMS.**—The disease may set in with attacks of nausea and vomiting, spontaneous in character. Toward the close there may be pain with retraction of the abdomen, and even features suggestive of peritonitis. A marked anorexia may be present. The gastric symptoms are variable throughout the course; occasionally they are absent. Attacks of diarrhœa are frequent and come on without obvious cause.

(c) **ASTHENIA**, the most characteristic feature, may be manifested early as a feeling of inability to carry on the ordinary occupation, or the patient may complain constantly of feeling tired. There may be an extreme degree of muscular prostration in an individual apparently well nourished, whose muscles feel firm and hard. The cardio-vascular asthenia is manifest in a feeble, irregular action of the heart, which may come on in paroxysms, in attacks of vertigo, or syncope, in one of which the disease may prove fatal. The blood pressure is low, falling to 70 or 80 mm. of Hg.

Headache is a frequent symptom; convulsions occasionally occur. Pain in the back may be an early and important symptom. Anæmia, specially referred to by Addison, is not common. In a majority of the patients the

## DISEASES OF THE GLANDS OF INTERNAL SECRETION

blood count is normal. The sugar content of the blood has been found to be low in some cases. McMunn described an increase in the urinary pigments, and a pigment has been isolated of very much the same character as the melanin of the skin.

The termination is by syncope, which may occur early in the disease, by gradual progressive asthenia, or by the development of tuberculous lesions. A noisy delirium with urgent dyspnoea may precede the fatal event.

**Diagnosis.**—Pigmentation of the skin is not confined to Addison's disease. The following conditions may give rise to an increase in the pigment; some of which, e. g., *a* and *b*, are due, as in Addison's disease, to disturbance in the chromaffin system.

(a) *Abdominal growths*—tubercle, cancer, or lymphoma. In tuberculosis of the peritoneum pigmentation is not uncommon.

(b) *Pregnancy*, in which the discoloration is usually limited to the face. Uterine disease is a common cause of a patchy melasma.

(c) *Hæmochromatosis*, associated with cirrhosis of the liver, pigmentation of the skin, and diabetes.

(d) In overworked persons of constipated habit there may be a patchy staining of the face and forehead.

(e) The vagabond's discoloration, caused by the irritation of lice and dirt, may reach a high grade and has been mistaken for Addison's disease.

(f) In rare instances there is deep discoloration of the skin in melanotic cancer, so general that it has been confounded with *melasma suprarenale*.

(g) In certain cases of *exophthalmic goitre* abnormal pigmentation occurs.

(h) In a few rare instances the pigmentation in *scleroderma* may be general and deep.

(i) In the face there may be an extraordinary degree of pigmentation due to innumerable small black comedones. If not seen in a very good light, the face may suggest argyria. Pigmentation of an advanced grade may occur in chronic ulcer of the stomach and in dilatation of the organ.

(j) *Argyria* has sometimes been mistaken for Addison's disease.

(k) *Arsenic* may cause a most intense pigmentation of the skin.

(l) With arterio-sclerosis and chronic heart disease there may be marked melanoderma.

(m) In *pernicious anæmia* the pigmentation may be extreme, most commonly due to the prolonged administration of arsenic.

(n) There is a form of deep pigmentation, usually in women, which persists for years without any special impairment of health. The pigmentation is a little more leaden than is usual in Addison's disease.

(o) In *ochronosis* there may be a deep melanotic pigmentation of the face and hands.

(p) In von Recklinghausen's disease the pigmentation may be uniform and suggestive of adrenal disease.

In any case of unusual pigmentation these conditions must be excluded; the diagnosis of Addison's disease is scarcely justifiable without the asthenia. In many instances it is difficult early in the disease to arrive at a definite conclusion. The syncope, low blood pressure, nausea, and gastric irritability are important. As the lesion of the capsules is almost always tuberculous, in doubtful cases the tuberculin test may be used.

**Prognosis.**—The disease is usually fatal. The cases in which pigmentation is slight or does not occur run a more rapid course. There are occasionally acute cases which, with great weakness, vomiting, and diarrhoea, prove fatal in a few weeks. In a few cases the disease is much prolonged, even to six or ten years. In rare instances recovery has taken place, and periods of improvement, lasting many months, may occur.

**Treatment.**—When asthenia appears the patient should be confined to bed and sudden efforts and muscular exercise should not be allowed. Fatal syncope may occur at any time. For the debility arsenic and strychnia are useful; for the diarrhoea large doses of bismuth, and for the irritability of the stomach very simple diet and alkalies. The diet should be light and nutritious; sugar should be given freely. As the disease is nearly always tuberculous an open air treatment may be carried out. Tuberculin may be tried cautiously, particularly if the patient is seen early. If there is any evidence of syphilis, specific treatment should be given cautiously.

Operation has been suggested and should be considered if there is a tumor in the situation of the adrenals. In any case, unless there were supernumerary adrenals and a considerable portion of the extra-capsular chromaffin intact, the operation would be useless.

**ADRENAL THERAPY.**—The relation of Addison's disease to the adrenals is not the same as that of myxedema to the thyroid gland, in which the insufficiency is relieved by the administration of thyroid preparations. The tuberculous nature of the lesions in most cases of Addison's disease is an obstacle, and there is usually widespread involvement of the sympathetic system. A large series of cases have been treated with various preparations, but only a very few with satisfactory results. In only three of our patients was there marked improvement. In one, all the severe symptoms disappeared, the pigmentation cleared, and the patient died subsequently of an acute infection. The adrenals were found sclerotic but not tuberculous. The dried gland may be given in doses of from 5 to 20 grains (0.3 to 1.3 gm.) three times a day. Epinephrine by injection may be tried also.

## II. OTHER AFFECTIONS OF THE SUPRARENAL GLANDS

**Lesions of the Adrenal Cortex.**—Remarkable changes in the secondary sexual characters have been associated with tumors and other lesions of this part—the so-called suprarenal genital syndrome. Pseudo-hermaphroditism has been found in connection with hyperplasia of the cortex, as in a case in which the internal organs were those of a female but the external had male characters. The reverse may occur. Premature puberty, with the development of the secondary sexual characters, obesity and overgrowth of hair, may appear as early as the fifth or sixth year. After puberty the presence of a tumor may lead to the condition known in women as virilismus or hirsutismus, in which a growth of hair occurs on the face, the voice becomes masculine, and the muscular strength may increase. Later, as the signs of tumor develop, there are emaciation, pigmentation, and mental changes.

**Hyper- and Hypo-function of the Adrenals.**—The state of our knowledge is far too uncertain to make it worth while to discuss the clinical vagaries grouped under the terms hyper- and hypo-epinephrinæmia. In acute adrenal

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insufficiency there may be abdominal pain with diffuse tenderness, marked prostration, malaise, vomiting, a small soft pulse, low blood pressure, hypoglycæmia and sometimes diarrhoea, convulsions, coma and death. The suggestion of Sergent that the vaso-constrictor skin reflex, causing the "white line," is an evidence of adrenal insufficiency has not been supported, but in some cases of asthenia and low blood pressure the response to the administration of adrenal gland substance is prompt. That certain disturbances come under these headings can not be doubted but much work and many observations are necessary before they can be accurately stated.

**Hæmorrhage.**—Acute hæmorrhagic adrenalitis presents a picture somewhat like acute pancreatitis—a sudden onset with pain, vomiting, profound prostration and death within a few days. In other cases convulsions occur or a typhoid state with profound asthenia results. In children the disease may be associated with purpura, both cutaneous and visceral.

**Tumors.**—Orthmann (1921) analyzed 147 operative and 140 post mortem cases of tumor of which 66 were carcinoma, 55 sarcoma and 115 hypernephroma. Stevens (1923) collected 75 cases of adeno-carcinoma of the gland of which one-third were in infants or young children. Metastases occurred early and were usually widespread. Weakness, gastro-intestinal disturbance and pain were the commonest symptoms. The pain usually extended up to the corresponding shoulder and across the abdomen anteriorly. Pigmentation, hæmaturia (possibly due to pressure on the renal vein) and overgrowth of hair were found occasionally. Successful removal has been done. In children excessive genital development with overgrowth of hair and obesity has been found, as noted by Bullock and Sequeira, who collected a number of cases. On this account a suggestion has been made that the adrenal cortex has an hormonal internal secretion which influences sexual development. Robert Hutchison described a remarkable syndrome in children of adrenal tumor, exophthalmos, and cranial tumors; and William Pepper (tertius) described a form characterized by rapid growth, diffuse involvement of the liver, and great distention of the abdomen without ascites or jaundice.

**Carotid Glands.**—Situated at the bifurcation of the carotid arteries, these bodies, each about the size of a grain of wheat, belong to the chromaffin group. Their function is unknown. They are of interest as the seat of tumors, benign at first but which may become malignant. The situation at the bifurcation of the carotid artery, movable laterally but not vertically, single, smooth, not tender or painful, a transmitted pulsation with a thrill and a murmur, bulging of the wall of the pharynx, slow growth and long duration, are the important features.

## II. DISEASES OF THE THYMUS GLAND

The thymus in structure has little resemblance to the other ductless glands, with the exception of the epiphysis cerebri, and must be classed as an epithelial rather than as a lymphoid organ. At birth the thymus gland weighs about 12 grams; from the first to the fifth year about 23 grams; from the sixth to the tenth year about 26 grams; from the eleventh to the fifteenth year about 37½ grams, and from the sixteenth to the twentieth year about 25½ grams,

## DISEASES OF THE THYMUS GLAND

after which it undergoes a gradual atrophy (Hammar). Involution not taking place, a "persistent thymus" remains.

The function of the gland is not known. There is an obscure relationship between the thymus and the sexual glands. After castration N. Patton found persistency and hypertrophy of the gland. A disturbance of the normal development of the bones, particularly in ossification, also occurs (Basch) and there is an increase in the excitability of the nerves. The nature of the internal secretion is unknown. Many experiments have been made with extract from the gland, but without definite results.

### I. HYPERTROPHY OF THE THYMUS

The size of the gland varies so greatly that it is not easy to define the limits between persistency and enlargement. Between the manubrium sterni and the vertebral column in an infant of eight months the distance is only 2.2 cm. (Jacobi), so that it is easy to understand how an enlarged gland may induce what Warthin calls "thymic tracheostenosis." There would appear to be, as this author suggests, three groups of cases:

(a) *Thymic stridor*, either congenital or developing soon after birth, varying in intensity and aggravated by crying and coughing.

(b) *Thymic asthma*, sometimes known as Kopp's or Miller's asthma, is an exaggerated and more persistent form of the stridor. While much dispute exists as to this form, there can be no doubt as to its occurrence, as there are cases in which complete relief has followed removal of the gland. Olivier collected 39 cases of thymectomy with 24 recoveries.

(c) Lastly, in some cases sudden death has occurred, usually in connection with the condition of lymphatism about to be described.

Persistence of the gland has been met with in many affections, such as Graves' disease, Addison's disease, acromegaly, myasthenia gravis, rickets, etc. Many observers regard the association of an enlargement with Graves' disease as more than accidental and as a sort of compensatory process.

### II. ATROPHY OF THE THYMUS

This is met with accidentally in children who show no special pathological changes, especially as Ruhräh has shown, in marasmus and the chronic wasting disorders of children. Of other morbid conditions, hæmorrhages are not uncommon. Inflammation of the gland (thymitis) may occur by infection from neighboring tissues. Mediastinal tumors may originate in the remnants of the thymus; dermoid tumors and cysts have also been met with; tuberculosis and syphilis of the gland are occasionally seen. The condition described by Dubois in congenital syphilis, in which there are fissure-like cavities in the gland filled with a purulent fluid, is probably post mortem softening.

### III. STATUS THYMICO-LYMPHATICUS: LYMPHATISM

**Definition.**—A combination of constitutional anomalies among which are hyperplasia of the lymphoid tissues and of the thymus, hypoplasia of the cardio-vascular system, and peculiarities of configuration.

## DISEASES OF THE GLANDS OF INTERNAL SECRETION

Formerly the condition was regarded as specially important in young children, but it is found both in children and adults. In Bellevue Hospital, 457 cases were found among 5,652 autopsies (8 per cent.). Of these only 92 were below the age of twenty years (Symmers). In young adults lymphatism is common. Borst and Grace found lymphatic hyperplasia in 56 per cent. of 2,000 men killed in action and in 86 per cent. of those aged 19 and 20.

The results of the condition are various; among them are: (1) The liability to sudden death. This may be from several causes. (a) Anaphylaxis. Necrosis occurs in the lymphoid tissues with resulting sensitization. With further necrosis a fatal attack may result. (b) Cerebral hæmorrhage. The hypoplastic arteries rupture easily, as from slight trauma, which is a point of medico-legal importance. (c) In young children sudden death may result from pressure of the enlarged gland ("thymic death"), but this is probably very rare. (2) Increased susceptibility to acute infections and decreased resistance to them. This applies particularly to endocarditis, pneumonia, cerebro-spinal fever and sepsis. (3) In women there is increased danger in childbirth. (4) Psychical instability. The subjects form a considerable proportion of cases of drug addiction and suicide.

**Pathology.**—Symmers describes two forms—status lymphaticus and recessive status lymphaticus. The former shows well-developed changes in the lymphoid tissues and occurs at an age when these structures are active. The recessive form shows atrophic changes in the lymphoid structures which vary with the time of involution. Of 219 cases, 118 were instances of status lymphaticus 89 of the recessive form and 42 were border-line cases, tending toward recession. In the status lymphaticus form the thymus was hyperplastic, the average weight being about 25 gm. No instance was found of death being due to pressure from the thymus. Histologically the thymus showed hyperplasia, which may be extreme. Necrotic changes were marked in the lymph nodes, especially in the cases of sudden death from slight causes. This is regarded by Symmers as being in close relation to anaphylaxis.

**Symptoms.**—Children with lymphatism are often fat, may be anæmic and flabby but are usually regarded as in good health. The tonsils are enlarged and adenoids are present. They have little resistance to infections and are easily upset by trifling ailments. They are often subject to nasal catarrh, mouth breathing is common, and vaso-motor changes are frequent. The blood may show a marked lymphocytosis. The enlarged thymus may be shown by dulness over the upper sternum and to each side of it which shifts upward with extreme retraction of the head (Boggs). There may be bulging or the gland may be felt in the episternal notch. The X-ray shadow may be distinct. In these cases there may be attacks, often after a fit of temper or a crying spell, in which the child shows noisy breathing, stridor and cyanosis. Respiration may stop for some seconds or death may occur.

After puberty the condition is easily recognized. In males the main points are: (1) A slender thorax, rounded arms and thighs, and a suggestion of the feminine type. (2) A soft delicate skin. (3) A scanty growth of hair on the face, especially on the upper lip and chin, and in the axillæ, with the pubic hair showing the feminine distribution. (4) The external genitals may be poorly developed; some are cryptorchids. (5) The cervical and axillary glands may be palpable. In females the main features are: (1) A slender

thorax and extremities. (2) A soft delicate skin. (3) Scanty axillary and pubic hair. (4) Hypoplasia of the genital organs.

**Diagnosis.**—Suspected cases should be carefully examined before trifling operations. The enlargement of the superficial glands, of the tonsillar tissues and of the spleen is easily determined. The adult forms are readily recognized from the general characteristics.

**Treatment.**—In children it is well to reduce the sugar and starch in the diet to a minimum, giving skim milk, eggs, meat, green vegetables and fruits. A general tonic treatment with iron and arsenic should be given. A large thymus causing compression may require removal but treatment by the X-ray is often successful. In the adult forms there is no special treatment.

#### IV. TUMORS OF THE THYMUS GLAND

These are rare and apparently there are less than 100 cases on record, the majority being sarcomata. Symmers and Vance divide tumors of the thymic parenchyma into those originating from the predominant cell of the adult thymus, the lymphocyte, and those from the early predominant epithelial cell. The latter are very rare and occur in patients over 50. The lymphocytic tumors occur earlier in life; many are in children. The symptoms are usually those of pressure and are variable. Early diagnosis, especially aided by X-rays, gives a chance of removal. Otherwise X-ray therapy is indicated.

### III. DISEASES OF THE THYROID GLAND

#### I. CONGESTION

At puberty, in girls, often at the onset of menstruation, the gland enlarges; in certain women the neck becomes fuller at each menstruation, and it was an old idea that the gland enlarged at or after defloration. From mechanical causes, as tight collars or repeated crying, the gland may swell for a short time. Slight enlargement is common in acute infections.

#### II. THYROIDITIS

**Etiology.**—Inflammation of the gland, which is nearly always secondary to some infection, may be simple or purulent. It occurs with typhoid fever, small-pox, measles, pneumonia, rheumatic fever, mumps, influenza, and after tonsillectomy. Epidemics of thyroiditis have been reported. It is a rare disease in ordinary hospital practice.

**Symptoms.**—The whole gland may be involved or only one lobe. There are swelling, pain on pressure, redness over the affected part, and, when suppuration occurs, softening or fluctuation. The pressure signs may be acute and severe. In suppurative cases the tenderness is marked and the diagnosis from diffuse cellulitis may be difficult. Often the acute inflammation subsides spontaneously. Myxœdema has followed destruction of the entire gland by acute suppuration.

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**Treatment.**—Sedatives may be required for discomfort. Iodide should not be used in any way. An ice-bag should be applied locally. With signs of suppuration free drainage should be secured.

There is a *primary chronic thyroiditis* (also termed sclerotic or woody) which usually occurs in males, between the ages of 20 and 40, without previous thyroid disease. There is inflammation, with the formation of dense connective tissue which involves the deep tissues of the neck and may compress the trachea and recurrent laryngeal nerves. Sections usually show very little thyroid tissue. The onset is with swelling of the neck, which increases rapidly; the mass is hard, smooth, and firmly fixed to the deeper structures. There is rarely pain or tenderness. Owing to the hardness of the mass, it may be regarded as malignant. Spontaneous disappearance has occurred. If treatment is necessary on account of pressure on the trachea, the least possible interference is wise as myxœdema has followed removal of the slight remaining thyroid tissue and treatment by X-rays.

### III. TUMORS OF THE THYROID

Of these the most important are: (a) *Infective granulomata*—tuberculosis, actinomycosis, and syphilis, which are rare. Tuberculosis may be mistaken for exophthalmic goitre. Swelling of the gland may occur in recent syphilitic infection, and gummata in the congenital form. (b) *Malignant tumors* are more common than statistics usually indicate. Wilson, in a study from the Mayo Clinic, states that among 16,558 thyroid operations, 207 cases of malignant disease were found; in addition there were 83 inoperable cases. Carcinoma is much more common than sarcoma. It is more frequent in women and occurs especially in the fifth decade. Adenomas play an important part in the etiology and so the history of recent growth in a previous nodular tumor is frequent. In a certain number the growth is slow. There is usually a progressive increase in the size of the thyroid, which is often irregular. With this, signs of pressure on the larynx, trachea, and œsophagus are common. Pain is not uncommon and there may be a good deal of cardio-vascular disturbance. In later stages the tumor is large and the surface veins are distended. The skin may become adherent and the pressure signs are marked. Erosion of the trachea or œsophagus may occur. Metastases are common, especially in the lungs. The *diagnosis* has to be made from adenoma and operation should be done if there is any doubt. *Treatment* is surgical and should not be delayed. The use of radium or X-rays is advisable after operation and as a palliative measure.

### IV. ABERRANT AND ACCESSORY THYROIDS

In various places, from the root of the tongue to the arch of the aorta, fragments of thyroidal tissue have been found. These aberrant portions of the gland are very apt to enlarge and undergo cystic degeneration. In the mediastinum they may form large tumors, and in the pleura an accessory cystic thyroid may occupy the upper portion, and a case was reported by F. A. Packard, in which the cystic gland filled nearly the entire side. The so-called lingual thyroid is not uncommon, varying in size from a hemp seed

## DISEASES OF THE THYROID GLAND

to a pea, usually free in the deep muscles of the tongue, or attached to the hyoid bone. When enlarged the *lingual goitre* may form a tumor of considerable size. The true thyroid gland has been absent, and removal of the lingual goitre has been followed by myxœdema.

### V. SIMPLE GOITRE

**Definition.**—A chronic enlargement of the thyroid gland, due to lack of iodine, occurring sporadically or endemically.

**Distribution.**—Goitre is widely distributed throughout the United States and Canada. In England it is common in certain regions; the Thames valley, the Dales, Derbyshire, Sussex, and Hampshire. It is very prevalent about Oxford and the upper Thames valley. In Switzerland, in the mountains of Germany and Austria, the mountainous districts of France, and in the Pyrenees the disease is very prevalent. In regions of Central Asia, in the Alysinnian mountains, and in the Himalayas there are many foci of the disease.

**Etiology.**—The disease is rarely congenital except in very goitrous districts. Cases are most common at or about puberty, and the tendency diminishes after the twentieth year. Women are much more frequently attacked than men, in a proportion of 6 or 8 to 1. The disease occurs at every latitude and in every altitude, in valleys and in plains, and in various climates. It seems to be less prevalent by the seashore.

In the diffuse colloid goitre of adolescence there is probably exhaustion of the thyroxin in the tissues with a resulting stimulation of the thyroid. The formation of thyroxin is disturbed from *shortage of iodine*. Infection may influence the exhaustion of thyroxin in the tissues. It is suggested that bacteria in the digestive tract may influence the absorption of iodine. The essential cause is lack of a sufficient amount of iodine.

**Morbid Anatomy.**—Usually the whole gland is involved, but one lobe only may be attacked. There are various changes which may be combined: (1) Parenchymatous, in which there is a general increase in the thyroid tissue; (2) colloid, with a great increase in the amount of colloid; (3) cystic, in which the cysts contain colloid material; hæmorrhage may occur into the cysts; and (4) fibrous, with a great increase in fibrous tissue. There is an increase in the colloid material of the follicles. Degenerations of various kinds are common, particularly cystic, in which there are many large and small cavities with colloid contents. In some of these cystic forms there are papillary ingrowths into the alveoli. Sometimes extensive hæmorrhages occur in the gland.

**Symptoms.**—The increase in size is usually insidious and in many cases there are no symptoms and the only complaint is of the disfigurement from the enlargement. Symptoms are due to pressure; the trachea may be flattened from pressure, usually from an enlarged isthmus, or narrowed by circular compression. The symptoms are more or less marked stridor and cough, which may persist for years without special aggravation. They may be present with very large glands, with the small encircling goitre, or with a goitre which passes deeply beneath the sternum. Pressure on the recurrent nerves may cause attacks of dyspnoea, particularly at night, and the voice may be

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altered. Pressure on the vagus is not common. Sometimes there is difficulty in swallowing, and the veins of the neck may be compressed. The heart may be involved, from pressure on the vagi. Other cardiac disturbances probably mean hyperthyroidism. The basal metabolism may be below normal (10 to 15 per cent.). This is altered very rapidly by giving thyroxin. The *diagnosis* of enlargement of the gland rarely gives difficulty unless it is intrathoracic. Care should always be taken to recognize hyperthyroidism. With *intrathoracic* goitre there are dyspnoea and choking sensations, especially with movement, and the patient sleeps with the head high. Dulness, decreased movement of the larynx on swallowing, laryngeal paralysis, venous obstruction, and signs of bronchial compression may be found. The X-ray study is important. Sometimes the tumor may appear and disappear.

**Prognosis.**—Many cases in the young get well; too often the tumor persists, but it may disappear on a change of locality.

**Prevention.**—The brilliant work of Marine and his associates has shown that in a great majority of cases simple goitre can be prevented by the administration of iodine. Amounts of 5 grains (0.3 gm.) of the iodide given twice weekly for a month twice a year are usually sufficient.

**Treatment.**—Iodine in some form is used extensively, and often is curative. Its affect is to stimulate the gland to healthy action. In young people 2 to 5 grains (0.13 to 0.3 gm.) of potassium or sodium iodide may be given daily. Iodine injections into the gland are not advisable. Iodine may be applied externally as an ointment (5 per cent.). The X-rays have been tried with success. When the gland is large, surgical measures are indicated.

### VI. HYPOTHYROIDISM

#### (*Cretinism and Myxœdema*)

**Definition.**—A constitutional affection due to insufficiency of the thyroid gland, characterized by a myxœdematous condition of the subcutaneous tissues and mental disturbance, and anatomically by atrophy of the thyroid gland.

**History.**—As early as 1859 Schiff had noted that in the dog removal of the gland was followed by certain symptoms. Gull described "a cretinoid change in women," and in the eighties the observations of Ord and other English physicians separated a well defined entity called "myxœdema."

Kocher (in 1883) reported that 30 of his first 100 thyroidectomies had been followed by a very characteristic picture, to which he gave the name "cachexia strumipriva," an observation already made in the previous year by the Reverdins, who had recognized the relation of this change to the disease known as "myxœdema." The researches of Horsley, and the investigation of the Committee of the Clinical Society of London, made it clear that the changes following complete removal of the gland, cachexia strumipriva, myxœdema, and sporadic cretinism, were one and the same disease, due to insufficiency of the thyroid gland. Schiff and Horsley demonstrated that animals could be saved by the transplantation of the glands. Lastly came the discovery of George Murray and Howitz that feeding with thyroid extract replaced the gland function, and cured the disease. The first patient given thyroid by Murray in 1891 died in 1919, aged 74, from heart disease. The activity of the

gland is connected with the metabolism of iodine, of which the maximum amount in the gland does not exceed 25 milligrams.

Kendall isolated the active principle which he terms *thyroxin* and which contains 65 per cent. of iodine. It is an amino-acid which enters into reaction and is regenerated so that it can repeat the process. It acts as does thyroid extract in myxœdema. There is a quantitative relation between thyroxin and the rate of basal metabolism.

The outcome of a host of researches has been the recognition of the enormous importance of the internal secretion of the gland, which is essential for normal growth in childhood, and has a marked influence on metabolism. It stimulates both vegetative nervous systems.

**Etiology.**—This is clear in the cases of complete removal of the gland or when atrophy of the remaining portion of the gland occurs. Probably atrophy is responsible in the cases occurring after exophthalmic goitre and infections. In cretinism usually one or both parents have thyroid disease. In some cases the gland is absent. In myxœdema a number of factors may be operative: (1) at the *menopause* it is not uncommon; (2) with frequent childbearing and sometimes with pregnancy; (3) following acute infections; (4) with other endocrine disturbances; and (5) in old age. Females are much more often affected than males and especially between 40 and 50.

**Clinical Forms.**—There are three groups of cases—cretinism, myxœdema proper, and operative myxœdema. To Felix Semon is due the credit of recognizing that these are one and the same condition and all due to loss of function of the thyroid gland.

**CRETINISM.**—Two forms are recognized—the sporadic and the endemic. In the *sporadic* form the gland may be congenitally absent, or is atrophied after one of the specific fevers, or the condition develops with goitre. The disease is not very uncommon; the histories of 58 cases were collected in a few years in the United States and Canada (Osler). It is more common in females than in males—35 in the series.

**Morbid Anatomy.**—Absence of the gland or complete fibrous atrophy is the common condition. Goitre with any trace of gland tissue is rare. In the sporadic form sometimes the hypophysis and thymus have been found enlarged. Arrest of development, a brachycephalic skull in the endemic, and a dolichocephalic in the sporadic form, are the chief skeletal changes.

**Symptoms.**—Hypothyroidism should be suspected in all children overweight at birth, nine pounds or over. In such children signs of delayed bone development should be looked for by an X-ray study. Otherwise the condition is rarely recognized before the child is six months old and often not until later. These children usually lose weight from malnutrition; by the age of two years they are under size and under weight. The development of bone is retarded. It is noticed the child does not grow so rapidly and is not bright mentally. The tongue looks large and hangs out of the mouth. The hair may be thin and the skin very dry. Usually by the end of the first year and during the second year the signs become very marked. The face is large and bloated, the eyelids are puffy and swollen; the *ala nasi* are thick, the nose is depressed and flat. Dentition is delayed, and the teeth which appear decay early. The abdomen is swollen, the legs are thick and short, and the hands and feet are undeveloped and pudgy. The face is pale and sometimes has a

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waxy, sallow tint. The fontanelles remain open; there is muscular weakness, and the child can not support itself. In the supraclavicular regions there are large pads of fat. The child does not develop mentally and may lapse into imbecility.

In cases in which the atrophy of the gland follows a fever the condition may not come on until the fourth or fifth year, or later. This is really, as Parker determined, a *juvenile myxœdema*. In a few of the sporadic forms cretinism develops with an existing goitre. It may retard development, bodily and mental, without ever progressing to complete imbecility.

**ENDEMIC CRETINISM.**—This occurs wherever goitre is very prevalent, as in parts of Switzerland, Savoy, Tyrol, and the Pyrenees. It formerly prevailed in parts of England. The clinical features are the same as in the sporadic form, stunted growth and feeble mind, plus goitre. Whatever the causal agent may be, it is the interference with the function of the gland that leads to the cretinous changes.

The *diagnosis* in a typical instance is very easy after one has seen a case or good illustrations. Infants a year or so old sometimes become flabby, lose their vivacity, or show a protuberant abdomen, and lax skin with slight cretinoid appearance. These milder forms are probably due to transient functional disturbance in the gland. Other causes of defective development, especially syphilis, should be excluded. It may be difficult to determine the basal metabolism but the effect of giving thyroid extract is of great aid in diagnosis. The *prognosis* depends on the time when treatment is begun. If this is early the child may become normal. In older children there may be improvement but a normal condition does not follow often. Cretins usually die young from intercurrent disease.

**MYXŒDEMA OF ADULTS (*Gull's Disease*).**—Women are much more frequently affected than men—in a ratio of 6 to 1. The disease may affect several members of a family, and it may be transmitted through the mother. In some instances there has been first the appearance of exophthalmic goitre. The symptoms of myxœdema may disappear during pregnancy or may develop post partum. Myxœdema and exophthalmic goitre may occur in sisters. Extreme forms are not so common in America as in England. C. P. Howard collected 100 American cases, of which 86 were in women.

The *onset* is usually insidious with mental dulness and loss of memory. Obscure pains, especially in the legs, are common; they are aggravated by rubbing or pressure. The pain is situated in the skin and subcutaneous tissues and is marked on pinching the skin. In other cases, due to involvement of the ligaments, there may be relaxation of the joints, causing pain and conditions such as flat-foot or lordosis. The clinical features are marked increase in the general bulk of the body, a firm, inelastic swelling of the skin, which does not pit on pressure; dryness and roughness, which tend with the swelling to obliterate in the face the lines of expression; imperfect nutrition of the hair; local tumefaction of the skin and subcutaneous tissues, particularly in the supraclavicular region. Perspiration is usually much decreased. The physiognomy is altered in a remarkable way: the features are coarse and broad, the lips thick, the nostrils broad and thick, and the mouth enlarged. The hair of the eyebrows is often scanty, especially on the outer part. Over the cheeks, sometimes the nose, there is a reddish patch. There is a striking

slowness of thought and of movement. The memory becomes defective, the patients grow irritable and suspicious, and there may be headache. In some instances there are delusions and hallucinations, leading to a final condition of dementia. The gait is heavy and slow. The temperature may be below normal and the patients often suffer in cold weather. The pulse is usually slow. Constipation is generally a marked feature. Hæmorrhage sometimes occurs, probably due to decreased coagulability of the blood. There is often a moderate secondary anæmia with a normal leucocyte count. Albuminuria is sometimes present, more rarely glycosuria. The tendon reflexes give a slow movement and greater stimulation than normal is needed to elicit a response. Death is usually due to some intercurrent disease, most frequently tuberculosis (Greenfield). The thyroid gland is diminished in size and may become completely atrophied and converted into a fibrous mass. The subcutaneous fat is abundant, and in one or two instances a great increase in the mucin has been found. The larynx is also involved.

The basal metabolism is reduced 20 to 40 per cent. below the normal.

The course is slow but progressive, and extends over ten or fifteen years. A condition of acute and temporary myxœdema may develop with enlargement of the thyroid and may follow exophthalmic goitre. The symptoms of the two diseases have been combined. In one case a young man increased in weight enormously during three months, then had tachycardia with tremor and active delirium and died within six months of the onset.

OPERATIVE MYXŒDEMA; CACHEXIA STRUMIPRIVA.—Horsley showed that complete removal of the thyroid in monkeys was followed by the production of a condition similar to that of myxœdema, sometimes associated with spasms or tetanoid contractures, and followed by apathy and coma. An identical condition sometimes follows extirpation of the thyroid in man. If a small fragment of the thyroid remains, or if there are accessory glands, the symptoms do not develop. Operative myxœdema is very rare in America. Some years ago only two cases were found, one of which, McGraw's, referred to in previous editions of this work, has been cured.

The *diagnosis* of marked myxœdema is easy, as a rule. The general aspect of the patient—the subcutaneous swelling and the pallor—suggests nephritis, which may be strengthened by the discovery of casts and albumin in the urine; but the solid character of the swelling, the exceeding dryness of the skin, the yellowish white color, the low temperature, the loss of hair, the dull, listless mental state and the low basal metabolism should differentiate the conditions. In dubious cases not too much stress should be laid upon the supraclavicular swellings. There may be marked fibro-fatty enlargements in this situation in healthy persons, the supraclavicular pseudo-lipomata of Verneuil. In mild cases the diagnosis is often overlooked because myxœdema is not considered. The basal metabolism and the results of thyroid therapy are important.

Jelliffe points out that in some cases *tabes dorsalis* may be diagnosed owing to the pains, unequal and sluggish pupils, myotonia, and decreased patellar and tendon Achilles reflexes.

Hypothyroidism should be considered in children who are dull and backward, in women who have symptoms suggesting a premature menopause, in obesity, and in those with constipation the cause for which is obscure.

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**Treatment.**—The patients suffer in cold and improve greatly in warm weather. They should therefore be kept at an even temperature, and should, if possible, move to a warm climate during the winter months. Repeated warm baths with massage are useful. Our art has made no more brilliant advance than in the cure of the disorders due to insufficiency of the thyroid gland. That we can to-day rescue children otherwise doomed to helpless idiocy—that we can restore to life the hopeless victims of myxœdema—is a triumph of experimental medicine for which we are indebted very largely to Victor Horsley and his pupil Murray. Transplantation of the gland was first tried; then Murray used an extract subcutaneously. Hector Mackenzie in London and Howitz in Copenhagen introduced the method of feeding. The gland is efficacious in a majority of all the cases of myxœdema in infants or adults. It makes little difference how it is administered. The dried gland is the most convenient. It is well to begin with the *Thyroideum siccum* U. S. P. 1 grain (0.065 gm.) three times a day. The dose may be increased gradually until the proper dose for each patient is found. Care should be taken to be sure of the strength of the preparation which is given. In many cases there are no unpleasant symptoms; in others there are irritation of the skin, restlessness, rapid pulse, delirium and in rare instances ionic spasms, the condition to which the term *thyroidism* is applied. The results, as a rule, are unparalleled by anything in the whole range of curative measures. Within six weeks a poor, feeble-minded, toad-like caricature of humanity may be restored to mental and bodily health. Loss of weight is one of the first and most striking effects; one patient lost over 30 pounds within six weeks. The skin becomes moist, the urine is increased, the perspiration returns, the temperature rises, the pulse rate quickens, the mental torpor lessens and the basal metabolism increases. Ill effects are rare.

The treatment, as Murray suggests, must be carried out in two stages—one, early, in which full doses are given until the cure is effected; the other, the permanent use of small doses sufficient to preserve the normal metabolism. In the cases of cretinism it seems necessary to keep up the treatment indefinitely as relapse may follow the cessation of the use of the extract.

### VII. ADENOMATOUS GOITRE

In the human thyroid there are usually groups of undeveloped thyroid cells; with the need for increased activity these “rests” grow and form the adenomata very often present in simple adolescent goitre. They are very variable in size and number. The frequent association of colloid and adenomatous goitre is to be noted. The simple adenomatous goitre may be diffuse or nodular. It usually develops about or soon after puberty and may persist indefinitely without causing symptoms. In other cases toxic symptoms may appear years later (toxic adenomatous goitre). Probably a variety of causes may be responsible, such as nervous disturbance and infections.

In *simple adenomatous goitre* the picture is very much the same as in simple goitre but the gland may be more irregular and nodular in places.

In *toxic or hyperfunctioning adenomatous goitre* (secondary hyperthyroidism) the condition of hyperthyroidism is added. As a rule this happens only after many years of enlargement of the gland and so the patients are

usually forty years of age or over. The onset of symptoms is usually gradual and for a time they may not be marked. *Fatigue* is of special importance. The clinical features are much like those of Graves' disease with certain differences. There is exophthalmos but the other eye signs are lacking. Tremor may be absent or coarse. The tachycardia is not so marked and is more often influenced by sleep or digitalis. Hypertension (especially an increase in the diastolic pressure) and evidence of myocardial degeneration (auricular fibrillation) are more common. Vaso-motor disturbances are less marked. The thyroid gland is large and often irregular; it does not often show pulsation, thrill or murmur. The psychical features are less marked and loss in weight is gradual. The basal metabolism is increased.

In **diagnosis**, the history of goitre for many years and the clinical features are usually sufficient, particular importance belonging to increased basal metabolism, which is often the deciding factor. The distinction from "neurasthenic" conditions may be difficult and often depends more on a careful study than on any special test, except basal metabolism. As patients with toxic goitre are resistant to the usual effects of quinine, 10 grain (0.6 gm.) doses may be given four times a day for several days. The absence of any of the usual results suggests hyperthyroidism.

The **prognosis** is variable and depends on the severity of the condition and the evidence of myocardial damage. It is essentially a chronic condition.

The **prevention** is that of simple goitre and it is to be hoped that by the administration of iodine or iodide many cases can be prevented.

The **treatment** is much like that of Graves' disease. Special attention should be given to any foci of infection. Special interest attaches to X-ray treatment which in many cases is successful. As the results of surgical treatment are usually excellent, it should be advised if other measures are not promptly successful. The presence of considerable myocardial change is not a necessary contra-indication.

## VIII. HYPERTHYROIDISM; EXOPHTHALMIC GOITRE

(*Graves', Basedow's, or Parry's Disease*)

**Definition.**—A disease characterized by goitre, exophthalmos, tachycardia, and tremor, associated with a perverted or hyperactive state of the thyroid gland and increased activity of the vegetative nervous system.

The essential nature of Graves' disease is in doubt. The thyroid changes, which represent extreme stimulation of the gland, may be entirely *secondary* and result from various causes. It may be a question of too much thyroxin production or the formation of an abnormal substance, which Plummer suggests may be an incomplete thyroxin molecule. The disturbed function of the thyroid may be associated with a lack of iodine. Excessive administration of thyroid extract does not cause exophthalmic goitre. A distinction should be made between hyperthyroidism and Graves' disease. Not all the cases of over-activity of the gland go on to exophthalmic goitre, but it is probable that the possibility of this progress exists. It may be difficult to classify some of the borderline cases.

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**Historical Note.**—In the posthumous writings of Caleb Hillier Parry (1825) is a description of 8 cases of Enlargement of the Thyroid Gland in Connection with Enlargement or Palpitation of the Heart. In the first case, seen in 1786, he also described the exophthalmos: "The eyes were protruded from their sockets, and the countenance exhibited an appearance of agitation and distress, especially in any muscular movement." The Italians claim that Flajani described the disease in 1800. Moebius states that his original account is meagre and inaccurate, and bears no comparison with that of Parry. If the name of any physician is to be associated with the disease, undoubtedly it should be that of the distinguished old Batn physician. Graves described the disease in 1835 and Basedow in 1840.

**Etiology.**—*Age.*—In Sattler's collection of 3,477 cases only 184 were under the age of sixteen. *Sex.*—The proportion of females is greatly in excess; in Sattler's collected cases the ratio was 5.4 to 1.

The exciting causes are probably varied and in many cases two or more are concerned. The acute infections, focal infections, previous thyroid disease and particularly nervous disturbances are factors. The influence of worry, anxiety, mental shock and severe fright is evident in many cases. In some patients a psychical factor seems to be sufficient and this is regarded by some as the usual cause.

A strong *family predisposition* may exist and five or six members may be affected.

**Pathology.**—The essential change consists in increased activity of the gland, which enlarges as a result of hyperplasia and shows increased vascularity. The normal colloid is greatly reduced or absent. The epithelial cells of the follicles show proliferation and the lymph-adenoid tissue is increased. These changes may occur only in limited areas of the gland tissue. The enlargement occasionally results in mechanical disturbance. The increased secretion causes definite results: (1) There is a great increase in metabolism; (2) other endocrine glands are affected, and (3) the sympathetic nervous system is stimulated. The active principle—thyroxin—has been isolated by Kendall. In many cases there is enlargement of the thymus, which may play a part in the lymphocytosis usually found (30-60 per cent.) with decrease in the neutrophils. Myxœdema may develop in the late stages, and there are transient cedema and in a few cases scleroderma, which indicate that the nutrition of the skin is involved. The degree of hyperthyroidism and dysthyroidism may be difficult to determine. The relative amounts of each may vary. The response to iodine therapy depends on the influence on dysthyroidism.

In rare instances the thyroid gland has been stated to be normal. In the majority of cases there is active hyperplasia of the gland, with enlarged and newly formed follicles, and an increase in the lymphoid tissue of the gland stroma. Involutionary and regressive changes are common; the hyperplasia may cease and the gland returns to the colloid state. Finally, in certain cases, atrophy of the cell elements takes place. The iodine content of the gland bears a direct relationship to the amount of colloid; the gland in hyperplasia has the lowest percentage, the pure colloid glands the highest.

**Symptoms.**—Acute and chronic forms may be recognized. In the acute form the disease may arise with great rapidity. In a patient of J. H. Lloyd's,

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of Philadelphia, a woman, aged thirty-nine, who had been considered perfectly healthy, but whose friends had noticed that for some time her eyes looked rather large, was suddenly seized with intense vomiting and diarrhoea, rapid action of the heart, and great throbbing of the arteries. The eyes were prominent and the thyroid gland was much enlarged and soft. The gastro-intestinal symptoms continued, the pulse became more rapid, the vomiting was incessant, and the patient died on the third day of the illness. The acute cases show marked toxæmia but are not always associated with delirium.

More frequently the onset is gradual and the disease is chronic. Toxic symptoms usually appear within a year. There are five principal features—tachycardia, exophthalmos, enlargement of the thyroid, tremor and increased basal metabolism.

**TACHYCARDIA.**—Rapid heart action is the most constant phenomenon. The pulse rate at first may be not more than 95 or 100, but when the disease is established it may be from 140 to 160, or even higher. The increase is most marked in the sympathicotonic cases. Irregularity is not common, except toward the close and may be due to auricular fibrillation. Auricular flutter may occur. In a well developed case the visible area of cardiac pulsation is much increased and the action is heaving and forcible. There is marked visible pulsation in the arteries. The capillary pulse is readily seen, and there are few diseases in which one may see at times with greater distinctness the venous pulse in the veins of the hands. The throbbing pulsation may be felt even in the finger tips. Vascular erythema is common—the face and neck are flushed and there may be a widespread erythema of the body and limbs. The blood pressure varies; there may be an early stage of hypertension followed by a long period of hypotension. In some patients hypertension may return after a time. Murmurs are usually heard, a loud apex systolic and at the base. The heart sounds may be very intense. In rare instances they may be heard at some distance from the patient; according to Graves, as far as four feet. Attacks of acute dilatation may occur with dyspnoea, cough, and a frothy bloody expectoration.

**EXOPHTHALMOS.**—A characteristic facial aspect is given by the staring expression, caused in part by protrusion of the eyeballs, but more particularly by retraction of the lids exposing the sclerae. The exophthalmos, which may be unilateral, usually follows the vascular disturbance. The protrusion may become very great and the eye may even be dislocated from the socket, or both eyes may be destroyed by panophthalmitis. The vision is normal. Graefe noted that when the eyeball is moved downward the upper lid does not follow it as in health. This is known as Graefe's sign. The palpebral aperture is wider than in health, owing to spasm or retraction of the upper lid. The patient winks less frequently than in health (Stellwag's sign). There is marked tremor of the lids and they contract spasmodically in advance of the elevating eyeball. Moebius called attention to disturbance of convergence of the two eyes. The majority of the eye signs are autonomic in origin. Changes in the pupils and in the optic nerves are rare. Pulsation of the retinal arteries is common.

**ENLARGEMENT OF THE THYROID** is the rule but may be absent. It may be general or in only one lobe, and is rarely so large as in ordinary goitre. The swelling is firm, but elastic. There are rarely pressure signs. The vessels

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is usually much dilated, and the whole gland may pulsate. A thrill may be felt and a systolic murmur heard. A double murmur is common and is pathognomonic (Guttman).

TREMOR was really first described by Basedow. It is involuntary, fine and about eight to the second. It is of great importance in the diagnosis of the early cases.

The *basal metabolism* (minimal heat production) shows a marked increase and this is an important aid in diagnosis. In very severe cases the increase may be 75 per cent. or over, in severe cases 50 to 75 per cent., and in milder forms from 20 to 50 per cent. Boothby in a study of 2,332 cases of exophthalmic goitre found in 94 per cent. a basal metabolism of +20 or over, in 5 per cent. between +11 and +20 and in 1 per cent. normal. (In 1,425 patients with adenoma with hyperthyroidism, 68 per cent. were +20 or over and 32 per cent. +11 to +20; in adenoma without hyperthyroidism the findings were within normal limits in all.)

Other features are anæmia, emaciation and slight fever. The blood shows lymphocytosis. Attacks of vomiting and diarrhoea may occur; the latter may be severe and distressing, recurring at intervals and without evident cause. Great complaint is made of the forcible throbbing in the arteries, often accompanied with unpleasant flushes of heat and profuse perspirations. An erythematous flushing is common. Pruritus may be a severe and persistent symptom. Multiple telangiectases have been described. Solid, infiltrated œdema is not uncommon and may be transitory. A remarkable myxœdematous state may supervene. Pigmentary changes are common and may be patchy or generalized. The coexistence of scleroderma and Graves' disease has been frequently noticed. Irritability of temper, change in disposition, and great mental depression occur. There is often marked emotional disturbance. An important complication is acute mania, in which the patient may die in a few days. Weakness of the muscles is not uncommon, particularly a feeling of "giving way" of the legs. If the patient holds the head down and is asked to look up without raising the head, the forehead remains smooth and is not wrinkled, as in a normal individual (Joffroy). A feature noted by Charcot is a great diminution in the electrical resistance, which may be due to the saturation of the skin with moisture owing to the vaso-motor dilatation (Hirt). Bryson noted that the chest expansion may be greatly diminished. The emaciation may be extreme. Glycosuria and albuminuria are not infrequent and true diabetes may occur. In women the menstrual function may be disturbed. The influence of pregnancy on the disease is variable.

The course is usually chronic, lasting several years, and often with remissions. After persisting for six months or a year the symptoms may disappear. There are remarkable instances in which the symptoms have come on with great intensity, following fright, and have disappeared again in a few days.

**Prognosis.**—Statistics are misleading as only the severe cases come under hospital treatment. Probably 65 per cent. of the patients make a good recovery and 10 to 12 per cent. die. Certain of the others go on to some degree of chronic invalidism. If rest and care do not influence the symptoms the outlook is serious as also in those in whom the disease begins after the age

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of 45. There are often periods of remission. In the hands of competent surgeons the mortality from operation is low and the results are excellent.

**Diagnosis.**—The typical cases are easily recognized but the difficulty comes with the partially developed forms and hyperthyroidism. The patient should be kept at rest and carefully studied. If the giving of thyroid extract (gr. i-ii, 0.06-0.12 gm.) for a few days increases the symptoms and pulse rate, it is significant. In acute cases syphilis should be excluded. The test of Goetsch which consists in the response to the injection of epinephrine (0.5 c. c.) is sometimes of value. An increase in the pulse rate and blood-pressure and aggravation of the general symptoms are the important points. It may aid in the diagnosis from early tuberculosis which may show features suggestive of hyperthyroidism. The response to quinine hydrobromide may be of value: 10 grains (0.6 gm.) are given three times a day and usually within a few days the normal individual has symptoms of cinchonism. They do not appear in exophthalmic goitre. Certain signs should suggest the possibility of hyperthyroidism: (1) tachycardia, (2) rapid emaciation without evident cause, (3) diarrhoea without evident cause, (4) lymphocytosis, and (5) nervous disturbance otherwise difficult to explain. Diagnosis in the borderline cases depends more on careful study and observation than on any single test. The most important point is the increase in basal metabolism, but this should be done by a competent worker and the necessity of repeated tests realized. An increase of at least 15 per cent. is necessary to have significance.

There is a group in which many of the features of Graves' disease are present but the basal metabolism is normal. Such cases represent "*autonomic imbalance*" (List, Hyman and Kessel). The most frequent feature is tachycardia, next coming thyroid enlargement. Tremor and eye signs are less common. A normal basal metabolism is the important diagnostic point.

**Treatment.**—It is usually well to try medical treatment before surgery is considered. Halfway measures should not be considered; the patient should be in bed, at absolute rest and excitement and irritation avoided. Any causes of worry should be corrected if possible. Long hours of sleep should be secured by sedatives if necessary. Any focus of infection should be treated. Tobacco, alcohol, tea and coffee should be forbidden. The patients should be liberally fed and sufficient food given to equalize the increased metabolism. In the diet, milk, buttermilk and foods prepared with milk should figure largely. Cereals, eggs, butter, bread or toast, vegetables and fruits may be given. Meat broths and meat are not to be given; small amounts of chicken may be taken occasionally. Water should be taken freely, best as distilled water, but, if not available, boiled water. An ice-bag should be applied over the heart. Of internal remedies, belladonna and ergot seem helpful in some cases. Quinine hydrobromide (gr. 10, 0.6 gm. three times a day) is often useful. Iodine in the form of Lugol's solution (m x, 0.6 c. c.) is often useful in cases of dysthyroidism. There may be marked improvement with its use and a trial of it is advisable before operation. The application of the X-rays or radium has been successful in many cases.

**Surgical Treatment.**—Operation is indicated, (1) when there are compression signs, (2) when there is no gain under a proper trial of medical treatment and (3) when medical treatment causes improvement but there is not complete recovery. Severe toxæmia is usually a contra-indication to sur-

## DISEASES OF THE GLANDS OF INTERNAL SECRETION

**Tetany.** Removal of part of the thyroid gland offers the best hope of permanent cure. It is remarkable with what rapidity all the symptoms may disappear after partial thyroidectomy. A second operation may be necessary in severe cases. Ligature of the arteries may be enough. Excision of the superior cervical ganglia of the sympathetic has one beneficial result, viz., the production of slight ptosis, which obviates the staring character of the exophthalmos.

### IV. DISEASES OF THE PARATHYROID GLANDS

The parathyroid bodies occur, as a rule, in two pairs on either side of the lateral lobes of the thyroid gland; small ovoid structures from 6 to 8 mm. in length. They have an internal secretion supplementing that of the thyroid gland and influencing calcium metabolism. Following their removal in animals there are twitching spasms of the voluntary muscles, gradual paralysis with dyspnoea, and death from exhaustion. These sometimes disappear when an extract of the parathyroid is injected into a vein, or if the parathyroid glands are fed or transplanted. The association of tetany with disturbance of the function of the parathyroid seems definitely established. MacCallum has shown the importance of these glands in controlling calcium metabolism, and it is possible that in disturbance of this is to be sought the cause of the great excitability of the nervous system and of tetany.

These studies have thrown great light upon various spasmodic disorders of children, and some have gone so far as to embrace such conditions as laryngismus, infantile convulsions, and tetany under the term "spasmophilia." These glands have also hormonal relations, as yet not thoroughly understood, with the other ductless glands, and have some influence on carbohydrate metabolism. The association of the glands with tetany is sufficient warrant for treating this disease here, though it may result from other causes.

#### TETANY

**Definition.**—Hyperexcitability of the neuro-muscular system with bilateral chronic or intermittent spasms of the muscles of the extremities. There are definite changes in the calcium metabolism, possibly due to disturbance in the functions of the parathyroid glands.

**Etiology.**—It occurs in epidemic form, particularly in the spring, sometimes with slight fever and behaves like an acute infection. It may occur in, or follow, the infections, typhoid fever, measles, etc. Of 8 cases reported by C. P. Howard, 4 were associated with dilatation of the stomach, 2 with hyperacidity, 1 with chronic diarrhoea, and 1 with lactation. In adults the gastro-intestinal group is the most common. It may follow successive pregnancies—the "nurse's contracture" of Trousseau.

In children it is common with rickets and in gastro-intestinal affections of artificially fed infants associated with wasting. Laryngo-spasm and child crowing are usually manifestations of tetany. In infantile tetany hæmorrhages into the parathyroid glands have been found. Forced breathing, as in acute diseases, hysteria, early anaesthesia and with excessive exercise, may cause

## DISEASES OF THE PARATHYROID GLANDS

alkalosis and tetany follows. The same may result from over-dosage of sodium bicarbonate. There is disturbance of the acid-base equilibrium.

The fact that tetany may follow removal of the thyroid (*tetania strumipriva*) led to the experimental studies showing a supposed relationship of the disease to the parathyroid gland. Removal of these bodies is followed by tetany, and in animals transplantation of living parathyroids cures experimental tetany; indeed, there are cases of human tetany that have been cured by transplantation. Where no disease of the glands has been found a parathyroid insufficiency is assumed.

There is evidence that the function of the parathyroid glands is concerned with calcium metabolism or guanidin metabolism (Paton) or both. They may be concerned in the regulation of the acid-base equilibrium. That there is a striking reduction in the calcium content of the blood in tetany as shown by MacCallum has been confirmed by Howland and Marriott in clinical cases (falling from the normal 10 to 11 mgms. per 100 c. c. to an average of 5.6 mgms.); but these writers conclude that the cause of the calcium deficiency is not yet explained and that the parathyroid theory lacks confirmation.

**Morbid Anatomy.**—Atrophy, hemorrhages, adenomas, cysts and inflammations have been found in the parathyroids, but the glands have been found normal in fatal cases.

**Symptoms.**—The tonic spasms occur chiefly in the upper extremities; the arms are flexed across the chest with the hands in the so-called "obstetric" position, the proximal phalanges flexed, the middle and distal extended with the thumb contracted in the palm. The legs are extended with plantar flexion of the feet and toes. The muscles of the face are not so often involved, but there may be trismus and spasm of the muscles of expression. The spasms may last only for a few hours or the condition may persist for days or weeks, recurring in paroxysms. Contracture of the back muscles is rare; occasionally there are general convulsions. Laryngo-spasm may occur with noisy inspiration. Pain is variable, sometimes being very severe. The pulse may be quickened and the temperature raised. Disturbance of sensation is rare. In chronic cases, the skin looks tense or drawn, there may be oedema, the hair falls out, and the teeth may subsequently show defects in the enamel. Perinuclear cataract may follow a prolonged attack.

Certain additional features are present:

*Trousseau's sign* is thus described by him—"So long as the attack is not over, the paroxysm may be reproduced at will. This is effected by simply compressing the affected parts, either in the direction of their principal nerve trunks, or over their blood-vessels so as to impede the arterial or venous circulation." The spasm is really caused by pressure on the nerves. It may be elicited months, or even years, after an attack. It is not always present.

*Chvostek's phenomenon* depends on an increased excitability of the motor nerves. A slight tap on the facial will throw the muscles into spasm, sometimes only limited groups. It is sometimes seen in debilitated children who have not had tetany and not infrequently in healthy children.

*Erb's phenomenon* is due to increased electrical excitability of the motor nerves. In normal infants a cathodal opening contraction is not caused by a current of less than 5 milliamperes; contraction is obtained in tetany with

## DISEASES OF THE GLANDS OF INTERNAL SECRETION

much less. Anodal hyperexcitability is also present, especially in latent tetany, but it may occur in normal infants and in other conditions.

**Diagnosis.**—The disease is readily recognized. Between the attacks, or even long after, the signs may be obtained. The common carpopedal spasm of debilitated infants is regarded by some as mild tetany. The predisposing factors, gastro-intestinal disease, thyroidectomy, pregnancy, etc., should be borne in mind. There is rarely any difficulty in differentiating tetanus, epilepsy or functional cramps. Hysterical spasm may give difficulty.

**Prognosis.**—Post-operative cases may prove fatal. Death in the gastro-intestinal forms is usually from the primary conditions. Recovery is the rule in children. The cause is an important element in prognosis.

**Treatment.**—In children the condition with which the tetany is associated should be treated. Baths and cold sponging are recommended and often relieve the spasm as promptly as in child-crowing. Bromide of potassium and chloral hydrate may be tried. In severe cases chloroform inhalations may be given. Massage, electricity, and the spinal ice-bag have also been used with success. Cases, however, may resist all treatment, and the spasms recur for many years. Parathyroid gland should be given (gr. 1/10, 0.006 gm.) and the addition of thyroid is sometimes of use.

Calcium therapy has proved efficacious in doses of gr. v-xv (0.3-1 gm.) of the lactate every three or four hours. The symptoms are promptly relieved, but the drug must be continued for some weeks. Excessive alkaline therapy should be avoided. Ammonium chloride (gr. 30-90, 2-6 gm. a day) has proved useful, probably by correcting alkalosis.

In gastric tetany, especially when due to dilatation of the stomach, the mortality is high, and recovery without operative interference is rare. Regular, systematic lavage with large quantities of saline or mildly antiseptic solutions is sometimes beneficial.

## V. DISEASES OF THE PITUITARY BODY

The hypophysis cerebri consists of two lobes, (*a*) an anterior lobe, originating from the roof of the pharynx and composed of large granular epithelial cells arranged in columns surrounded by large venous spaces into which their secretion discharges; and (*b*) a smaller posterior lobe which arises from the floor of the third ventricle and is composed (1) of a central neuroglial portion (*pars nervosa*) and (2) an investment of epithelial cells (*pars intermedia*). The secretion of the posterior lobe is supposed by some to find its way into the cerebro-spinal fluid.

Complete experimental removal of the gland is fatal (Paulesco). Partial removal leads, in young animals, to a stunting of growth, to adiposity and failure of sexual development, in adult animals to adiposity and genital dystrophy (Cushing).

Modern knowledge of the functions of the gland began with the studies of Marie on its relation to acromegaly and gigantism. Then Schäfer and Oliver discovered that injection of an extract of the gland caused a rise in blood pressure. Since these observations an enormous amount of work has been done, and we now appreciate the remarkable influence of this small struc-

ture upon the processes of development and metabolism. Briefly, the anterior lobe influences growth and development, and is necessary to life; the posterior lobe influences the metabolism of the carbohydrates and fats.

Disturbances in the function of the pituitary gland are not clearly grouped into the effects of deficiency and excess, though one can differentiate states of hyper- and hypo- pituitarism. The hypophysis appears to be closely related to other glands of internal secretion and involvement of any member of the series causes a readjustment in the activity of the others. Owing to the situation of the gland it is very liable to feel the effect of pressure from neighboring or even distant lesions, so that disturbance of function may be due not only to a primary involvement, but to secondary compression. As a result of experimental work and clinical studies Cushing prefers to group the conditions associated with disturbance of the function of the gland under the term "dyspituitarism" and recognizes a number of groups:

(a) Cases of tumor growth showing signs of distortion of neighboring structures, and the constitutional effects of altered glandular activity. The X-rays show changes in the configuration of the pituitary fossa; there are pressure signs on the adjacent cranial nerves, bi-temporal hemianopia, optic atrophy, and oculomotor palsies. Uncinate fits are not unusual. The *convulsive seizures* due to hypopituitarism usually begin during adolescence and the patients show clinical features of under-activity of the gland with X-ray evidence of pituitary change. The giving of 5 to 10 grains (0.3 to 0.6 gm.) of the whole gland daily may be curative. There is a form of *headache* associated with pituitary disturbance.

(b) Cases in which the neighborhood manifestations are pronounced but the constitutional features are slight. The characteristic regional signs of tumor are marked, but there may be slight or very transient evidence of disturbed glandular activity, perhaps only disturbed carbohydrate metabolism with adiposity.

(c) Cases in which the neighborhood manifestations are absent or slight, though the glandular symptoms are unmistakable. The gland is not so large as to cause regional symptoms. There are skeletal changes either of over- or undergrowth. Disturbance of carbohydrate metabolism is a matter of modified posterior lobe activity, occurring as a lowering of the assimilation limit, often associated with the early stages of acromegaly, or a great increase in tolerance, as characterizes all grades of hypopituitarism. In posterior lobe insufficiency there is a tendency to the deposition of fat, subnormal temperature, drowsiness, slow pulse, dry skin, loss of hair, and an extraordinary high tolerance for sugars. Most cases of acromegaly fall in this group and show at first evidences of hyperpituitarism, and later of insufficiency. In the adult, adiposity, high sugar tolerance, subnormal temperature, psychic manifestations, and sexual infantilism of the reversive type indicate hypopituitarism and may exist without regional symptoms of tumor.

(d) Hypophysial symptoms may be shown by patients with internal hydrocephalus from any cause, probably by interference with the passage of the posterior lobe secretion into the cerebro-spinal fluid, and this obstructive dyspituitarism may result from any lesion, inflammatory or neoplastic, in the neighborhood of the third ventricle.

These are the most important of the groups but there are also cases with

## DISEASES OF THE GLANDS OF INTERNAL SECRETION

manifestations indicating involvement of other internal secretions with that of the hypophysis, and a large group in which transient hypophysial symptoms occur, as in pregnancy, cranial injuries and infectious diseases.

Disturbances in the function of the pituitary gland may lead to remarkable changes in growth; *hyperpituitarism* may lead to gigantism, when the process antedates ossification of the epiphyses—the Launois type; to acromegaly when it is of later date; *hypopituitarism* to adiposity, with skeletal and sexual infantilism when the process originates in childhood—the Fröhlich type; to adiposity and sexual infantilism of the reversible type when originating in the adult. Much has been done to clear the subject, but much remains, particularly to clear up the relations of the various types of infantilism—the Lorain, the Brissaud, the pancreatic, the intestinal—to the different internal secretions. One condition merits separate consideration, that differentiated by Marie and known as acromegaly.

### ACROMEGALY

**Definition.**—A dystrophy characterized by increase in size of the face and extremities associated with perverted function of the anterior lobe of the pituitary gland. The essence of the disease is a dystrophy of hypophysial origin (Marie), which, if it antedates ossification of the epiphyses, leads to gigantism, and in the adult leads to over-growth of the skeleton and other changes which we know as acromegaly.

**Etiology.**—It is a rare disease and rather more frequent in women. It affects particularly persons of large size. Twenty per cent. of acromegals are above six feet in height when the symptoms begin, and fully 40 per cent. of giants are acromegals (Sternberg). Trauma, the infections, and emotional shock have preceded the onset of the disease. The relation to the thyroid gland is emphasized by Anders and Jameson who found thyroid disturbance in about one-third of a series of 215 cases of acromegaly. Hypothyroidism is the most common change. They point out that the anterior lobe of the pituitary is supplied with sympathetic fibres while the posterior lobe is not.

**Pathology.**—Practically all of the cases show changes in the pituitary gland, hyperplasia, adenoma, fibroma, or sarcoma, causing distention of the sella turcica and, in the late stages, pressure on surrounding structures; the symptoms are in part due to disturbance of the function of the gland, and in part to the pressure on the adjacent parts.

The bones show the most striking changes; there is a general enlargement of the extremities, but the skeleton is more or less affected. The enlargement, due to a periosteal growth, is most evident in the hands and feet. The bones of the face are always involved. The orbital arches, frontal prominences, zygoma, malar, and nasal bones are increased in size; the lower jaw is elongated, thickened, and the teeth separated. The X-ray picture shows characteristic changes in the sella turcica. The skin and subcutaneous tissues are thickened and the hypertrophy is seen in the soft parts of the face as well. The brain has been found large, but the most important changes are those due to pressure at the base. The internal organs have been found enlarged, and in Osborne's case the heart weighed 2 lbs. 9 oz.

## DISEASES OF THE PITUITARY BODY

**Symptoms.**—When the pituitary gland is involved in tumor growth, which is common in acromegaly, the symptoms may be grouped into those due to mechanical effects and those associated with perversion of the secretion.

(a) **REGIONAL SYMPTOMS.**—Headache is common, usually frontal, and often very severe. Somnolence has been noted in many cases, and may be the first symptom. Ocular features occur in a large proportion of the cases, diminution of the fields of vision, bitemporal hemianopia, optic atrophy, and, in late stages, pressure on the third nerve and abducens. One eye only may be affected. Exophthalmos may occur. Deafness is not infrequent. Irritability, marked change in the disposition, great depression, and progressive dementia have been noted. Epistaxis and rhinorrhœa may be present.

(b) **SYMPTOMS DUE TO THE PERVERSION OF THE INTERNAL SECRETION** itself form the striking features of the disease. The patient's friends first notice a gradual increase in the features, which become heavy and thick; or the patient himself may notice that he takes a larger size of hat, or with the progressive enlargement of the hands a larger size of gloves. The enlargement of the extremities does not interfere with their free use.

The hypertrophy is general, involving all the tissues, and gives a curious spadelike character to the hands. The lines on the palms are much deepened. The wrists may be enlarged, but the arms are rarely affected. The feet are involved like the hands and are uniformly enlarged. The big toe, however, may be much larger in proportion. The nails are usually broad and large, but there is no curving, and the terminal phalanges are not bulbous. The joints may be painful and neuralgia is common. The head increases in volume, but not as much in proportion as the face, which becomes much elongated and enlarged in consequence of the increase in the size of the superior and inferior maxillary bones. The latter in particular increases greatly in size, and often projects below the upper jaw. The alveolar processes are widened and the teeth are often separated. The soft parts also increase in size, and the nostrils are large and broad. The eyelids are sometimes greatly thickened, and the ears enormously hypertrophied. The tongue in some instances becomes greatly enlarged. Late in the disease the spine may be affected and the back bowed—kyphosis. The bones of the thorax may slowly and progressively enlarge. With this gradual increase in size the skin of the hands and face may appear normal. Sometimes it is slightly altered in color, coarse, or flabby, but it has not the dry, harsh appearance of the skin in myxœdema. The muscles are sometimes wasted.

Also associated with disturbance of the function of the gland is glycosuria, noticed in many cases and common in the early stages; in the advanced stages there is an extraordinarily high tolerance for sugar. Symptoms on the part of other ductless glands are common. Goitre is of frequent occurrence. Myxœdema or a flabby obesity may occur late. Amenorrhœa is an early symptom in women. Impotence is common in advanced cases in men. The blood pressure and temperature are usually low.

**Diagnosis.**—This rarely offers difficulty; hypertrophic pulmonary osteoarthropathy and osteitis deformans are the conditions most likely to be confused. The general features and an X-ray study should prevent confusion.

**Treatment.**—The use of extracts of the gland has been extensively tried but with practically no results. Surgical treatment has been carried out in

## DISEASES OF THE GLANDS OF INTERNAL SECRETION

a number of cases, the chief indication being to give relief to the local pressure symptoms when there is marked glandular enlargement. Partial removal of the growth or the evacuation of a cyst may save the optic nerves from pressure atrophy. Sedatives have to be given for pain. Thyroid extract should be given if there is hypothyroidism.

### VI. DISEASES OF THE PINEAL GLAND

"That there is a small gland in the brain in which the soul exercises its functions more particularly than in the other parts" was the opinion of Descartes; and for more than two and a half centuries this was the type of our knowledge of the functions of the pineal gland. What we know now is derived chiefly from clinical cases. But the nature of the internal secretion is unknown; Barker believes that the pressure exerted by tumors of the gland may explain the symptoms. Some hold that disturbance of this gland may be associated with muscular dystrophy.

Disease of the gland, usually tumor, may cause (1) pressure symptoms, due to internal hydrocephalus, (2) focal symptoms, due to involvement of the cranial nerves, particularly those of the eyes, (3) features believed to be due to disturbance of the internal secretion, as sexual precocity, carbohydrate tolerance, obesity and increase in the growth of hair.

### VII. DISEASES OF THE SEX GLANDS

The endocrine part of the testicle is represented by the *interstitial cells* of Leydig and of the ovary by the interstitial cells and the cells of the corpus luteum. The secretions influence the development of the secondary sexual characters. *Hyperfunction* causes premature sexual development in both sexes. *Hypofunction* is shown (1) in eunuchs, in whom there is complete loss of the glands, and (2) in eunochoids, in whom there is insufficiency of the glands. In eunuchs there is lack of genital development, the body is large and fat, there is scanty growth of hair, and the psychical state is altered. In females whose ovaries are removed after puberty the features of the artificial menopause appear. *Eunochoids* differ according as the insufficiency occurred before or after puberty. There is usually involvement of other glands, especially the pituitary, with a pluriglandular syndrome. The individuals are usually tall and fat with absence of secondary sexual characters. The genitals show hypoplasia, and sterility, with disturbance of the sexual function, is the rule. In treatment, various combinations of glandular extracts may be used, especially testicular and pituitary in the male and ovarian in the female.

### VIII. INFANTILISM

**Definition.**—A disturbance in growth characterized by persistence of infantile or puerile characters and a general retardation of development, bodily and mental.

## INFANTILISM

**Etiology.**—It is not possible to make a satisfactory classification of the causes or of the cases of infantilism—in some no cause is evident, in others the failure in development has followed obvious disease, and there are cases directly dependent upon loss of some internal secretion.

**I. Cachectic infantilism** is by no means uncommon, as any serious chronic malady may delay sexual development. For example, the children affected with *hook-worm* disease may reach the age of 20 or older before puberty occurs. *Syphilis* is a very common cause. It occurs in cases of *chronic nephritis* beginning in childhood. In regions in which malaria is very prevalent delayed sexual development is not uncommon and we see it not infrequently in congenital heart disease. There is also a toxic infantilism due to the slow and prolonged action of alcohol and lead. The influence of lack of vitamins may be important.

**II. Idiopathic Infantilism (So-called Lornin Type).**—"In this variety the figure is so small that, at first sight, it looks like that of a child. When the patient is stripped, however, his outlines are seen to be those of an adult, and not those of childhood. The head is proportionately small, and the trunk well formed; for the shoulders are broad compared to the hips, and the bony prominences and the muscles stand out distinctly. We have before us a miniature man (or woman, as the case may be), and not one who has retained the characteristics of childhood beyond the proper time. There is, indeed, no growth of facial, pubic or axillary hair, yet the genital organs, though small, are well shaped and quite large enough for the size of the body. The intelligence in both sexes is generally normal" (John Thomson).

The cause is probably associated with perversion of the pituitary and thyroid secretions. It has been called an "angioplastic infantilism," in the belief that it was due to a defect of development of the vascular system.

**III. The Hormonic Type.**—Here we are on safer ground, as we know definitely of several varieties directly dependent upon disturbance of the internal secretions. The most important of these are:

(a) **THYROIDAL OR CRETINOID INFANTILISM.**—This has been described.

(b) **The FRÖHLICH TYPE, dystrophia adiposo-genitalis,** associated with a tumor of the pituitary region, is characterized by great obesity and genital hypoplasia. The symptoms are due to a secretory deficit, for they are capable of experimental reproduction by partial glandular extirpation in animals (Cushing). There are adult and infantile types, just as there are in myxedema; in the former the individual becomes fat and the sexual organs revert to the pre-adolescent state. The *Brissaud type* is in all probability due to dyspituitarism. A round, chubby face, under-developed skeleton, prominent abdomen, large layer of fat over the whole body, rudimentary sexual organs, no growth of hair except on the head, and absence of the second dentition, are some of the prominent features of this form, which Brissaud attributed to hypothyroidism, but which more likely is due to dyspituitarism.

(c) **PANCREATICO-INTESTINAL TYPE.**—Bramwell, Herter, Freedman, and others have reported cases of infantilism associated with intestinal changes. Bramwell thought the pancreas was at fault, and his patients improved under treatment with pancreatic extract. In Herter's case there were looseness of the bowels, often fatty stools, and a change in the flora of the intestine with a rise in the ethereal sulphates in the urine.

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**IV. Progeria.**—Under this term Hastings Gilford described a condition in children of incomplete development (infantilism) with premature decay. The facial appearance, the attitude, the loss of hair, wasting of the skin, are those of old age, and post mortem extensive fibroid changes are found, particularly in the arteries and kidneys. The condition is probably associated with unknown changes in the internal secretions.

## IX. DISEASES OF THE SPLEEN

### I. GENERAL REMARKS

Though a ductless gland, the spleen is not known to have an internal secretion, and its functions are ill understood. It is not an organ essential to life. In the fetus it takes part in the formation of the red blood corpuscles, and as it contains hamatoblasts, it is possible that in the adult this function may be exercised to some extent, particularly in cases of severe anæmia.

Hæmolysis is generally believed to be its special function, based upon the presence of a large percentage of organic compounds of iron, the deposit in the organ of blood pigments in various diseases, the presence of many macrophages containing red blood corpuscles, and upon the evidence, after removal of the spleen, of compensatory hæmolysis in many newly formed hæmo-lymph glands (Warthin).

Removal of the spleen, an operation practised by the ancients in the belief that it improved the wind of runners, is not, as a rule, followed by serious effects. There may be slight eosinophilia and temporary anæmia, and later there is usually slight leucocytosis, with relative increase of the lymphocytes. After removal, an accessory spleen may increase greatly in size. McLean and Craig reported a case of congenital absence of the spleen and found nine cases in which the individual reached middle life without disturbance. It may occur alone or with other defects.

In infections the organ enlarges and micro-organisms are present in large numbers. It has been supposed to play some part in the processes of immunity and phagocytosis goes on actively in the organ. In experimental anæmia caused by various hæmolytic agents the spleen enlarges, and in these conditions Bunting and Norris found evidence of vicarious blood formation. Chronic splenomegaly may be present with little disturbance of health.

### II. MOVABLE SPLEEN

Movable or wandering spleen is seen most frequently in women the subjects of enteroptosis. It may be present without signs of displacement of other organs. It may be found accidentally in individuals who present no symptoms whatever. In other cases there are dragging, uneasy feelings in the back and side. All grades are met with, from a spleen that can be felt completely below the margin of the ribs to a condition in which the tumor-mass impinges upon the pelvis; indeed the organ has been found in an inguinal hernia! In the large majority of all cases the spleen is enlarged. Sometimes it appears that the enlargement has caused relaxation of the ligaments; in

## DISEASES OF THE SPLEEN

other instances the relaxation seems congenital, as movable spleens have been found in different members of the same family. Possibly traumatism may account for some of the cases. Apart from the dragging, uneasy sensations and the worry in nervous patients, wandering spleen causes very few serious symptoms. *Torsion* of the pedicle may produce a serious condition, leading to great swelling of the organ, high fever, or necrosis. The diagnostic features are a sudden onset with intense pain and symptoms of peritonitis; a splenic tumor appears in a few hours with absence of the normal splenic dulness. There is absence of rigidity of the abdominal wall at first.

The *diagnosis* of a wandering spleen is usually easy unless the organ becomes fixed and is deformed by adhesions and perisplenitis. The shape and the sharp margin with the notches are the points to be specially noted.

The *treatment* is important. Occasionally the organ may be kept in position by a properly adapted belt and pad. Removal of the displaced organ has been carried out in many cases, but this is as a rule unnecessary. Splenectomy is indicated for torsion.

### III. RUPTURE OF THE SPLEEN

Special interest attaches to the rupture which may occur spontaneously or from trauma in typhoid fever or malaria. Rupture of a malarial spleen may follow a blow, a fall, or exploratory puncture. In India and in Mauritius rupture of the spleen is stated to be common. Fatal hæmorrhage may follow puncture of a swollen spleen with a hypodermic needle. Occasionally rupture results from the breaking of an infarct or abscess. The symptoms are those of hæmorrhage into the peritoneum, and the condition demands immediate laparotomy.

### IV. INFARCT, ABSCESS, CYSTS AND TUBERCULOSIS OF THE SPLEEN

Emboli in the splenic arteries causing *infarcts* may be infective or simple and are most frequent in ulcerative endocarditis and septic conditions. Infarcts may follow the formation of thrombi in the branches of the splenic artery in acute fevers, such as typhoid fever. In a few instances the infarcts have followed thrombosis in the splenic veins. They are chiefly of pathological interest. Infarct of the spleen may be suspected in endocarditis, septicæmia or pyæmia when there are pain in the splenic region, tenderness on pressure, and swelling of the organ; a well-marked friction rub is occasionally heard. Sometimes in the infective infarcts large *abscesses* are formed, and in rare instances the whole organ may be converted into a sac of pus.

*Tumors of the spleen*, *hydatid* and other *cysts*, abscess and *gummata* are rare conditions of anatomical interest. *Abscess* may result from extension but is usually due to hæmatogenous infection, especially in septic infection and typhoid fever. With the general features of infection there is a large tender spleen, often adherent to the abdominal wall. In *Hodgkin's disease* the organ may be enlarged and smooth, or irregular from the presence of nodular tumors.

*Cysts* are rare; the senior author saw but two, one an echinococcus, and the other a double cyst of the hilus. The latter probably arose from a hæma-

## DISEASES OF THE GLANDS OF INTERNAL SECRETION

oma, subcapsular or in the hilus. They have been successfully removed. Very small cysts occur in connection with polycystic disease of the liver and kidneys. A dermoid cyst has been described. The diagnosis of cysts is not often made; the mass is usually irregular in the region of the spleen, but the splenic outlines are marked. In the case with two cysts at the hilus, the tumor was very movable and irregular, and operation was urged on the grounds of mechanical discomfort and increase in size. Musser collected notes of 21 operations, all successful, in cysts of this sort.

*Primary tuberculosis* is rare. Winternitz collected 51 cases in 1912. In some cases the symptoms resemble those of an acute infection, with pain in the splenic region and enlargement of the organ. In the chronic cases there is progressive enlargement of the spleen, often with cyanosis and sometimes with polycythæmia. Splenectomy has been successful in some cases.

### V. PRIMARY SPLENOMEGALY WITH ANÆMIA

#### *(Splenic Anæmia, Banti's Disease)*

**Definition.**—A primary disease of the spleen of unknown origin, characterized by progressive enlargement, anæmia, a tendency to hæmorrhage, and in some cases a secondary cirrhosis of the liver, with jaundice and ascites. That the spleen itself is the seat of the disease is shown by the fact that complete recovery may follow its removal.

**History.**—The name "splenic anæmia" was applied to a group of cases by Griesinger in 1866. H. C. Wood, in 1871, described cases as the splenic form of pseudo-leukæmia. The real study of the disease was initiated by Banti in 1883. In France the condition was called "primitive splenomegaly," and many different types have been described. Here we deal with the form referred to in the definition as splenic anæmia and Banti's disease. There is unfortunately a confusion in terminology; some use the term Banti's disease as synonymous with splenic anæmia, while others restrict it to the cases of splenic anæmia in which cirrhosis of the liver develops; not all cases of splenic anæmia progress to the stage of Banti's disease. The latter usage is to be preferred.

**Etiology.**—In the majority of cases the enlargement of the spleen comes on without any recognizable cause. In a few cases malaria has been present, but in the greater number the first thing noticed has been the mechanical inconvenience of the big spleen. Males are more frequently attacked than females. It is a disease of young and middle life, the majority of cases occurring before the fortieth year. It is also met with in young children. Some of the cases of infantile splenic anæmia of von Jaksch and the Italian writers belong to this disease.

**Morbid Anatomy.**—The spleen is greatly enlarged, coming perhaps next to the size of the leukæmic organ. It is very firm, the capsule is thickened, the texture of the gland very tough and firm, and the whole in a state of advanced fibrosis. Banti described a proliferation of the endothelial cells of the venous sinuses of the pulp. The blood-vessels in the neighborhood of the spleen may be very large, particularly the vasa brevia, and the splenic vein itself and the portal vein may be enormously dilated, and show atheroma and

calcification. The lymphatic glands are not involved. Hyperplasia of the bone marrow has been found, but no other changes of special importance.

**Symptoms.**—The disease is extraordinarily chronic; eight of our cases had a longer duration than ten years. Usually the first feature is:

**Splenomegaly.**—The enlargement is uniform, smooth, painless, usually reaches to the navel, very often to the anterior superior spine, and the organ may occupy the whole of the left half of the abdomen. It may exist for years without any symptoms other than the inconvenience caused by the distention of the abdomen. Following an infarct pain may be present.

**Anæmia.**—Sooner or later the patients become anæmic. This develops with rapidity, and in children a severe and even fatal form may follow in a few weeks. More commonly the pallor is gradual and the patient may come under observation for the first time with swelling of the feet, shortness of breath, and the signs of advanced anæmia. The blood picture is that of a secondary anæmia with a low color index and a marked leucopenia. The red blood cells may fall as low as two million and in an average of a series of uncomplicated cases the leucocyte count was under 3,500 per c. mm. There are no special changes in the differential count. Following a severe hæmorrhage there may be a rise in the leucocytes. Some patients have permanent slight anæmia of the secondary type; others show recurring attacks of severe anæmia, which may be independent of hæmorrhage.

**Hæmorrhages.**—Bleeding, usually hamatemesis, may be a special feature and occur at intervals for many years. One of our patients had recurring attacks for twelve years, and one at the London Hospital for fifteen years (Hutchison). In such cases the diagnosis of ulcer of the stomach may be made. The bleeding may be of great severity. On several occasions one of our patients was brought into the hospital completely æsanguine; in two the hæmorrhage proved directly fatal; in a third the hæmorrhage proved fatal ten days after a successful splenectomy. The bleeding comes, as a rule, from œsophageal varices. Malena may be present. Hamaturia and purpura may occur.

**Ascites.**—Usually a terminal event, it may be due to the enlarged spleen itself or to secondary cirrhosis of the liver. When due to the liver, it may be associated with slight jaundice.

**Jaundice.**—Icterus has been rare in our cases. Enlargement of the spleen may persist for many years without any consecutive change in the liver. Slight jaundice may persist for years, sometimes with enlargement of the liver, in others with distinct reduction in its volume, and in either case with a progressive cirrhosis—the features to which Banti called special attention.

**Course.**—It is extraordinarily chronic. Some cases never progress to the stage of Banti's disease. A patient may for ten or twelve years have a large spleen causing no inconvenience, then an attack of anæmia may occur, from which recovery gradually takes place; or the first symptom may be ascites or a severe hæmorrhage from the stomach. As a rule, the anæmia becomes more or less chronic, with marked exacerbations, and in the later stages in many cases cirrhosis of the liver with jaundice and ascites develops.

**Diagnosis.**—Here may be mentioned other forms of splenomegaly.

**SPLENOmegaly WITH ACHOLURIC HÆMOLYTIC JAUNDICE.**—This form, first described by Minkowski and sometimes called after his name, is usually

## DISEASES OF THE GLANDS OF INTERNAL SECRETION

a familial form, often hereditary. It is consistent with good health throughout life, and there may be no symptoms. Characteristic features are: (a) its familial form; (b) chronic enlargement of the spleen; (c) good health; (d) chronic slight jaundice; (e) presence of urobilin in the urine, but absence of bile pigment. In a few instances gall stone colic has been present, due to small calculi. The red blood corpuscles have an increased fragility, the cause of which is unknown, but this is an essential feature. In the familial form good health is the rule, but in the acquired form the patient often becomes anæmic and is very ill.

Cures have been reported after splenectomy.

**SPLENOMEGALY OF THE GAUCHIER TYPE** (see page the c.

**SPLENOMEGALY WITH EOSINOPHILIA.**—This is a condition with marked enlargement of the spleen and leucocytosis. In some cases the leucocytes were 34,000 with 70 to 80 per cent. of eosinophils. Nucleated red cells are present in moderate numbers, both normoblasts and megaloblasts. The spleen showed a large number of eosinophiles. Following splenectomy there was a great increase in the number of leucocytes in two of the reported cases, to 138,000 and 211,000. The eosinophilia also increased to 90 per cent. in Giffin's case. It is suggested that there is a hyperplasia of certain elements in the bone marrow. Splenectomy in the reported cases seems to have resulted in benefit in the general condition but the eosinophilia became more marked. This may be due to the loss of the phagocytic influence of the spleen. The condition may represent an unusual form of leukaemia.

**SPLENOMEGALY WITH PRIMARY PYLETHROMBOSIS.**—Cases have been reported of enlarged spleen in connection with phlebitis of the splenic and portal veins, and such cases closely resemble splenic anæmia. The spleen is very large and there are jaundice and ascites with moderate anaemia. The recognition of the pylethrombosis is only made post mortem.

**HEPATIC SPLENOMEGALY.**—Three varieties of cirrhosis of the liver may lead to great enlargement of the spleen with anæmia and a symptom-complex resembling that of splenic anæmia.

(a) *Alcoholic Cirrhosis.*—With recurring hæmorrhages, a consecutive anæmia, ascites, and an unusually large spleen, the condition may simulate closely the last stage of splenic anæmia. The history, particularly the late appearance of the hepatic changes, may be the most important point. In the cases in which we have been in doubt the difficulty has arisen from an imperfect history and from the presence of recurring hæmorrhages.

(b) *Syphilitic Cirrhosis.*—Great enlargement of the spleen may occur with hepatic syphilis, congenital or acquired and the picture may be similar to splenic anæmia, slight jaundice, ascites, big spleen, recurring hæmorrhages, and marked anæmia. A search for the features of syphilis of the liver, the Wassermann test and the effect of syphilitic treatment are important but in some cases treatment has little effect. Syphilis may cause marked enlargement of the spleen without involvement of the liver.

(c) In a few cases of hypertrophic cirrhosis, as in Hanot's form and in hæmochromatosis, the spleen may be greatly enlarged, and when ascites and hæmorrhages occur, the clinical picture may be like that of splenic anæmia.

**SPLENOMEGALY IN PERNICIOUS ANÆMIA.**—Sometimes the spleen is greatly enlarged but the blood findings enable one to make the diagnosis.

## DISEASES OF THE SPLEEN

**TROPICAL SPLENOMEGALY.**—Kala-azar can be distinguished by the presence of the Leishman-Donovan bodies in the spleen. There are big spleens with anæmia in the Tropics which are not kala-azar, and the experience of some of the physicians in Cairo indicates that some of these are of the ordinary splenic anæmia type, in which removal of the organ cures the disease.

The cause of the enlarged spleen in malaria, leukæmia, and erythræmia is determined by the blood examination; in Hodgkin's disease, carcinoma, amyloid disease and infective endocarditis, other features usually prevent error.

**Treatment.**—There is only one means of radical cure—removal of the spleen. This should be done as early as possible, but if there is severe anæmia the usual treatment of this should be given and especially blood transfusion. When marked haemoglobin changes have occurred, operation is usually contra-indicated. In the latest far advanced for operation the treatment is that of any severe anæmia and with cirrhosis of the liver and ascites the usual measures should be adopted. If there is any evidence of syphilis active treatment for that should be given. The use of X-rays is of doubtful value but may be tried if operation is contra-indicated.

### VI. GAUCHER'S FORM OF SPLENOMEGALY (*Primary Endothelioma*)

This rare familial disease was described by Gaucher in 1882. Sappington (1920) in reporting a case found 24 in the literature. It usually begins in childhood before the 12th year, is more frequent in girls, and is characterized by the presence of large mononuclear cells of an endothelial type in the spleen, liver, lymph nodes and bone-marrow. The main features are a progressive enlargement of the spleen often to an enormous size, with the general health remaining good, followed by great enlargement of the liver without jaundice and rarely with ascites. There is a brownish yellow pigmentation of the skin, sometimes with wedge-shaped yellowish thickened areas on the conjunctiva on each side of the cornea. Anæmia usually appears late and is moderate; leucopenia is usual. The disease is chronic and may persist for twenty years or more without much disturbance of the general health. The special features usually give the diagnosis. Splenectomy is advisable if the general condition permits but it is not always curative.

## SECTION XII

# DISEASES OF THE NERVOUS SYSTEM

## A. GENERAL INTRODUCTION

**The Neurone.**—ITS STRUCTURE.—The nervous system is a combination of units called neurones, each composed of a receptive cell body and of conductors—namely, protoplasmic processes or dendrites, and the axis-cylinder process or axone. The dendrites conduct impulses toward the cell body (cellulipetal conduction) and the axones conduct them away from the cell (cellulifugal conduction). Depending upon whether the axones conduct impulses in a direction away from or toward the cerebrum they are called efferent or afferent. The axis-cylinder process gives off at varying intervals lateral branches called collaterals, running at right angles, and these, and finally the axis-cylinder process itself, split up into many fine fibres, forming the end brushes. These, known as arborizations, surround the body of one or more of the many other cells, or interlace with their protoplasmic processes. Thus, the terminals of the axone of one neurone are related to the dendrites and cell bodies of other neurones by contact or by concrescence.

**FUNCTION OF THE NEURONE.**—The function of the neurone is to conduct nervous impulses. Reduced to its simplest form, the mode of action may be represented by two cells, one of which, reacting to the environment, conducts impulses inward, whereas the other, awakened by this afferent impulse, conducts an impulse outward. This reflex response Marshall Hall showed to be the fundamental principle of action of the nervous system. The environment acts on the afferent neurones through special sense organs, so that a variety of afferent impulses, olfactory, visual, auditory, gustatory, tactile, painful, thermic, muscular, visceral and vascular may be originated. The efferent neurones convey impulses outward to non-nervous tissues, to the skeletal, visceral, and vascular muscles and to the secretory glands, whose activities are thus augmented or inhibited. The most important reflex centres lie in the bulbo-spinal axis. The situation of the vascular and respiratory centres in the bulb makes it the vital centre of the body. In the spinal cord the location of many reflex centres, particularly those for the muscle tendons and for some of the viscera, is represented in the table on page 921. The visceral mechanism is almost wholly regulated by the bulbo-spinal axis, and its reactions are usually unperceived. In conditions of disease the visceral reflexes may "rise into consciousness," and at such times referred pains and areas of tenderness are produced in the skin-fields of the spinal segments corresponding to the centre for registration of the visceral reflex.

**DEGENERATION AND REGENERATION OF THE NEURONE.**—The nutrition of the neurone depends upon the condition of the cell body, and this in turn upon the activity of the nucleus. If the cell is injured the process degen-

erates or the processes separated from the cell degenerate. Though the nerve cells cease to multiply soon after birth, they nevertheless retain remarkable powers of growth and repair. Injury to the cell body may not be recovered from, but if the axone be severed and degeneration take place in consequence, it may under favorable circumstances be replaced by sprouts from the central stump, and its function be regained. Even the peripheral section, independently of the cell body, may have the power of regeneration. It is probable that both factors play a part in the regeneration—namely, the down growth of the axone from the central end of the divided nerve as well as the changes in the periphery, most marked in the cells of the sheath of Schwann.

**Cell Systems.**—The cell bodies of the neurones are collected more or less closely together in the gray matter of the brain and spinal cord and in the ganglia of the peripheral nerves. Their processes, especially the axis-cylinder processes, run for the most part in the white tracts of the brain and spinal cord and in the peripheral nerves. In this way the different parts of the central nervous system are brought into relation with each other and with the rest of the body. Furthermore, the axis-cylinder processes arising from cells subserving similar functions are collected together into bundles or tracts, and though in many cases the course of these tracts and the functions which they possess are extremely complicated and as yet have not been completely unravelled, nevertheless some of them are simple and fairly well understood. By the study of degenerations resulting from injury or from the toxins of certain diseases which possess an affinity for one or another of these individual tracts or systems, it has been possible to trace the course of certain of them. Fortunately for the clinician, the best understood and the simplest system in its arrangement is that which conveys motor impulses from the cortex to the periphery—the so-called pyramidal tract.

**The Motor System.**—Motor impulses starting in the left side of the brain cause contractions of muscles on the right side of the body, and those from the right side of the brain in muscles of the left side of the body. Leaving out of consideration some few exceptions, it may be stated as a general rule that the motor path is crossed, and that the crossing takes place in the upper segment (Figs. 15 and 16). Every muscular movement, even the simplest, requires the activity of many neurones. In the production of each movement special neurones are brought into play in a definite combination, and acting in this combination specific movement is the result. In other words, all the movements of the body are represented in the central nervous system by combinations of neurones—that is, they are localized. Muscular movements are localized in every part of the motor path, so that in cases of disease of the nervous system a study of the motor defect often enables one to fix upon the site of the process, and it would be hard to over-estimate the importance of a thorough knowledge of such localization. A voluntary motor impulse starting from the brain cortex must pass through at least two neurones before it can reach the muscles, and we therefore speak of the motor tract as being composed of two segments—an upper and a lower.

**THE LOWER MOTOR SEGMENT.**—The neurones of the lower segment have the cell bodies and their protoplasmic processes in the different levels of the ventral horns of the spinal cord and in the motor nuclei of the cerebral nerves. The axis-cylinder processes of the lower motor neurones leave the spinal cord

## DISEASES OF THE NERVOUS SYSTEM

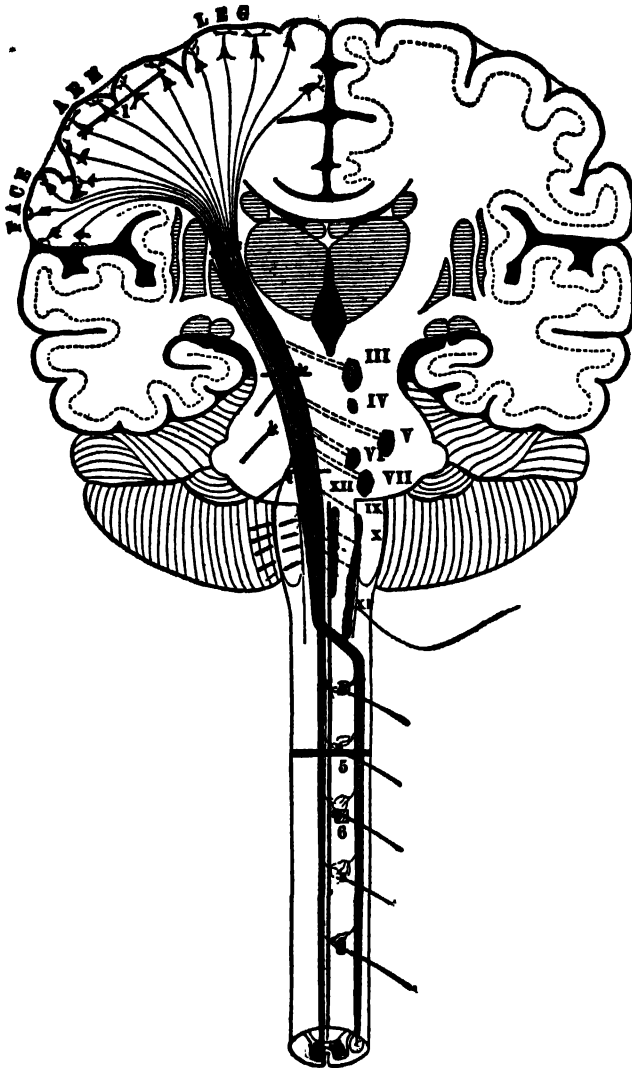


FIG. 15.—DIAGRAM OF MOTOR PATH FROM LEFT BRAIN.

The upper segment is black, the lower red. The nuclei of the motor cerebral nerves are shown in red on the right side; on the left side the cerebral nerves of that side are indicated. A lesion at 1 would cause upper segment paralysis in the arm of the opposite side—cerebral monoplegia; at 2, upper segment paralysis of the whole opposite side of the body—hemiplegia; at 3, upper segment paralysis of the opposite face, arm, and leg, and lower segment paralysis of the eye muscles on the same side—crossed paralysis; at 4, upper segment paralysis of opposite arm and leg, and lower segment paralysis of the face and the external rectus on the same side—crossed paralysis; at 5, upper segment paralysis of all muscles below lesion—spinal paraplegia; at 6, lower segment paralysis of muscles localized at seat of lesion—anterior poliomyelitis. (Van Gehuchten, modified.)

## GENERAL INTRODUCTION

in the ventral roots and run in the peripheral nerves, to be distributed to all the muscles of the body, where they end in arborizations in the motor end plates. These neurones are direct—their cell bodies and processes, and the muscles in which they end are all on the same side of the body.

The ventral roots of the spinal cord are collected, from above down, into small groups, which, after joining with the dorsal roots of the same level of the cord, leave the spinal canal between the vertebræ as the spinal nerves. That part of the cord from which the roots forming a single spinal nerve arise is called a segment, and corresponds to the nerve which arises from it and not to the vertebra to which it may be opposite. With the exception of

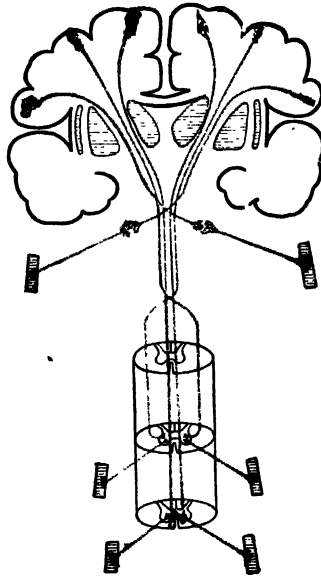


FIG. 16.—DIAGRAM OF MOTOR PATH FROM EACH HEMISPHERE, SHOWING THE CROSSING OF THE PATH, WHICH TAKES PLACE IN THE UPPER SEGMENT BOTH FOR THE CRANIAL AND SPINAL NERVES. (Van Gehuchten, colored.)

the cervical region, in which all the nerve roots but the eighth emerge from above the vertebræ, the roots of each segment for the remainder of the cord leave the spinal canal below the vertebra of corresponding number, and consequently, owing to the fact that during growth the bony canal lengthens much more than the cord itself, the more tailward one goes the greater is the discrepancy in position between each spinal segment and its particular vertebra. This must be borne in mind when determining upon the site of a lesion known to occupy a given segment, for it may lie far above the vertebra of like number and name. A chart has been prepared from numerous measurements by Reid showing the level of the various segments of the cord in relation to the spines of the vertebræ. The axis-cylinder processes which go to make up any one peripheral nerve do not necessarily arise from the same segment of the spinal cord; in fact, most peripheral nerves contain processes from several often quite widely separated segments. Most of the long-stripped

## DISEASES OF THE NERVOUS SYSTEM

muscles, furthermore, having originated in the embryo from more than one myotome, are innervated from more than one segment.

Our knowledge of the localization of the muscular movements in the gray matter of the lower motor segment is far from complete, but enough is known to aid materially in determining the site of a spinal lesion. The following table, in which is included for each of the spinal segments the centres of representation for the more important skeletal muscles, the main reflex centres, and the main location of the segmental skin-field, has been prepared from the studies of Starr, Edinger, Wichmann, Sherrington, Bolk, and others (pages 921, 922 and 923).

**THE UPPER MOTOR SEGMENT AND MOTOR AREAS OF THE CORTEX.**—The cell bodies of the upper motor neurones are found in the brain cortex lying for the most part in a strip anterior to the fissure of Rolando, and it is in this region that we find the movements of the body again represented.

True motor responses are elicited only by stimulation *anterior* to the Rolandic fissure; practically no point over the ascending frontal convolution fails to respond to stimulation. There is but slight extension of the motor cortex on to the paracentral lobule of the mesial surface of the brain. Movements are obtainable not only from the exposed part of the convolution, but also from its hidden surface to the very depths of the Rolandic sulcus. There is an area of representation for the trunk between the centres for the leg and arm, and also for the neck between those of the arm and face. The superior and inferior genua are the landmarks which indicate the situation of these small areas of representation for trunk and neck. These results have in large measure been confirmed by Cushing by unipolar electrical stimulation of the human cortex. From above down the motor areas occur in the following order: leg, trunk, arm, neck, head (Fig. 17). Those of the leg and arm occupy the upper half of the convolution, and that for the head, including movements of the face, jaws, tongue, and larynx, the lower half.

The speech centres are indicated in the diagram (Fig. 17) in accordance with the generally accepted views: that for motor speech occupies the posterior part of the left third frontal or Broca's convolution. It is a disputed point whether or not there is a separate centre presiding over the movements employed in writing. Some have assumed such a centre to be present in the second frontal convolution as indicated on the diagram. The conjugate movement of head and eyes to the opposite side has commonly been found in apes to follow stimulation of the external surface of the frontal lobe. Similarly movements of the eyes may be elicited from the occipital cortex, but probably none of these reactions are comparable to the more simple movements through the pyramidal tract which follow stimulation of the ascending frontal convolution.

The axis-cylinder processes of the upper motor neurones after leaving the gray matter of the motor cortex pass into the white matter of the brain and form part of the corona radiata. They converge and pass between the basal ganglia in the internal capsule. Here the motor axis-cylinders are collected into a compact bundle—the pyramidal tract—occupying the knee and anterior two-thirds of the posterior limb of the internal capsule. The order in which the movements of the opposite side of the body are represented at this level, as learned from experimental observations on apes, is given in Fig. 18.

# GENERAL INTRODUCTION

## LOCALIZATION OF THE FUNCTIONS IN THE SEGMENTS OF THE SPINAL CORD

SEGMENT.	STRIPED MUSCLES.	REFLEX.	SKIN-FIELDS (OF FIGS. 20 AND 21).
I, II and III C.	Splenius capitis. Hyoid muscles. Sterno-mastoid. Trapezius. Diaphragm (C III-V). Levator scapulæ (C III-V).	Hypochondrium (?). Sudden inspiration produced by sudden pressure beneath the lower border of ribs (diaphragmatic).	Back of head to vertex. Neck (upper part).
IV C.	Trapezius. Diaphragm. Levator scapulæ. Scaleni (C IV-T I). Teres minor. Supraspinatus. Rhomboid.	Dilatation of the pupil produced by irritation of neck. Reflex through the sympathetic (C IV-T I).	Neck (lower part to second rib). Upper shoulder.
V C.	Diaphragm. Teres minor. Supra- and infraspinatus (C V-VI). Rhomboid. Subscapularis. Deltoid. Biceps. Brachialis anticus. Supinator longus (C V-VII). Supinator brevis (C V-VII). Pectoralis (clavicular part). Serratus magnus.	Scapular (C V-T I). Irritation of skin over the scapula produces contraction of the scapular muscles. Supinator longus and biceps. Tapping their tendons produces flexion of forearm.	Outer side of shoulder and upper arm over deltoid region.
VI C.	Teres minor and major. Infraspinatus. Deltoid. Biceps. Brachialis anticus. Supinator longus. Supinator brevis. Pectoralis (clavicular part). Serratus magnus (C V-VIII). Coraco-brachialis. Pronator teres. Triceps (outer and long heads). Extensors of wrist (C VI-VIII).	Triceps. Tapping elbow tendon produces extension of forearm. Posterior wrist. Tapping tendons causes extension of hand (C VI-VII).	Outer side of forearm, front and back. Outer half of hand (?).
VII C.	Teres major. Subscapularis. Deltoid (posterior part). Pectoralis major (costal part). Pectoralis minor. Serratus magnus. Pronators of wrist. Triceps. Extensors of wrist and fingers. Flexors of wrist. Latissimus dorsi (C VI-VIII).	Scapulo-humeral. Tapping the inner lower edge of scapula causes adduction of the arm. Anterior wrist. Tapping anterior tendons causes flexion of wrist (C VII-VIII).	Inner side and back of arm and forearm. Radial half of the hand.

# DISEASES OF THE NERVOUS SYSTEM

## LOCALIZATION OF THE FUNCTIONS IN THE SEGMENTS OF THE SPINAL CORD (Continued)

SEGMENT.	STRIPED MUSCLES.	REFLEX.	SKIN-FIELDS (CF. FIGS. 20 AND 21).
VIII C.	Pectoralis major (costal part). Pronator quadratus. Flexors of wrist and fingers. Latissimus. Radial lumbricales and interossei.	Palmar. Stroking palm causes closure of fingers.	Forearm and hand, inner half.
I T.	Lumbricales and interossei. Thenar and hypothenar eminences (C VII-T I).		Upper arm, inner half.
II to XII T.	Muscles of back and abdomen. Erectores spinae (T I-IV). Intercostals (T I-T XII). Rectus abdominis (T V-T XII). External oblique (T V-XII). Internal oblique (T VII-L I). Transversalis (T VII-L I).	Epigastric. Tickling mammary region causes retraction of epigastrium (T IV-VII). Abdominal. Stroking side of abdomen causes retraction of belly (T IX-XII).	Skin of chest and abdomen in oblique dorso-ventral zones. The nipple lies between the zone of T IV and T V. The umbilicus lies in the field of T X.
I L.	Lower part of external and internal oblique and transversalis. Quadratus lumborum (L I-II). Cremaster. Psoas major and minor (?).	Cremasteric. Stroking inner thigh causes retraction of scrotum (L I-II).	Skin over lowest abdominal zone and groin.
II L.	Psoas major and minor. Iliacus. Pectineus. Sartorius (lower part). Flexors of knee (Remak). Adductor longus and brevis.		Front of thigh.
III L.	Sartorius (lower part). Adductors of thigh. Quadriceps femoris (L II-L IV). Inner rotators of thigh. Abductors of thigh.	Patellar tendon. Tapping tendon causes extension of leg. "Knee-jerk."	Front and inner side of thigh.
IV L.	Flexors of knee (Ferrier). Quadriceps femoris. Adductors of thigh. Abductors of thigh. Extensors of ankle (tibialis anticus). Glutei (medius and minor).	Gluteal. Stroking buttock causes dimpling in fold of buttock (L IV-V).	Mainly inner side of thigh and leg to ankle.
V L.	Flexors of knee (ham-string muscles) (L IV-S II). Outward rotators of thigh. Glutei. Flexors of ankle (gastrocnemius and soleus) (L IV-S II). Extensors of toes (L IV-S I). Peronæi.		Back of leg, and part of foot.

## GENERAL INTRODUCTION

### LOCALIZATION OF THE FUNCTIONS IN THE SEGMENTS OF THE SPINAL CORD (Continued)

SEGMENT.	STRIPED MUSCLES.	REFLEX.	SKIN-FIELDS (CF. FIGS. 20 AND 21).
I to II S.	Flexors of ankle (L V-S II). Long flexor of toes (L V-S II). Peronei. Intrinsic muscles of foot.	Foot reflex. Extension of Achilles tendon causes flexion of ankle (S I-II). Ankle-clonus. Plantar. Tickling sole of foot causes flexion of toes or extension of great toe and flexion of others.	Back of thigh, leg and foot; outer side.
III to V S.	Perineal muscles. Levator and sphincter ani (S I-III).	Vesical and anal reflexes.	Skin over sacrum and buttock. Anus. Perineum. Genitals.

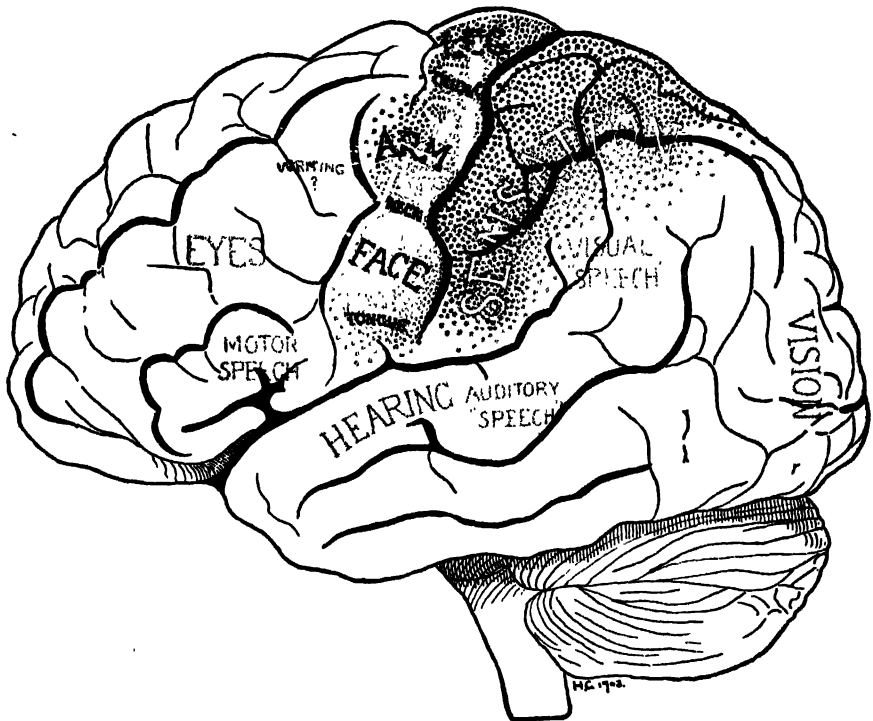


FIG. 17.—DIAGRAMMATIC REPRESENTATION OF CORTICAL LOCALIZATION IN THE LEFT HEMISPHERE, SHOWING THE SPEECH CENTRES.

The motor areas determined by unipolar faradic excitation of the anthropoid cortex (Sherrington and Grünbaum) are here shown stippled in red and lie anterior to the Rolandic fissure. The sensory areas presumably lie posterior to this fissure and are roughly indicated in blue without accurate delineation. Lying as it does on the upper surface of the hemisphere, the leg area should not be visible on a lateral view such as is given here.

## DISEASES OF THE NERVOUS SYSTEM

After passing through the internal capsule the fibres of the pyramidal tract leave the hemisphere by the crus, of which they occupy about the middle three-fifths. The movements of the tongue and lips are represented nearest the middle line.

As soon as the tract enters the crus, some of its axis-cylinder processes leave it and cross the middle line to end in arborizations about the ganglion cells in the nucleus of the third nerve on the opposite side; and in this way, as the pyramidal tract passes down, it gives off at different levels fibres which end in the nuclei of all the motor cerebral nerves on the opposite side of the body. Some fibres, however, go to the nuclei of the same side. From the crus the pyramidal tract runs through the pons and forms in the medulla oblongata the pyramid, which gives its name to the tract. At the lower part of the medulla, after the fibres going to the cerebral nerves have crossed the middle line, a large proportion of the remaining fibres cross, decussating with those from the opposite pyramid, and pass into the opposite side of the spinal cord, forming the crossed pyramidal tract of the lateral column (fasciculus cerebro-spinalis lateralis (Fig. 19, 1). The smaller number of fibres which do not at this time cross descend in the ventral column of the same side, forming the direct pyramidal tract, or Türcck's column (fasciculus cerebro-spinalis ventralis) (Fig. 19, 2).

At every level of the spinal cord axis-cylinder processes leave the crossed pyramidal tract to enter the ventral horns and end about the cell bodies of the lower motor neurones. The tract diminishes in size from above downward. The fibres of the direct pyramidal tract cross at different levels in the ventral white commissure, and also, it is believed, end about cells in the ventral horns on the opposite side of the cord. This tract usually ends about the middle of the thoracic region of the cord.

**The Sensory System.**—The path for sensory conduction is more complicated than the motor path, and in its simplest form is composed of at least three sets of neurones, one above the other. The cell bodies of the lowest neurones are in the ganglia on the dorsal roots of the spinal nerves and the ganglia of the sensory cerebral nerves. These ganglion cells have a special form, having apparently but a single process, which, soon after leaving the cell, divides in a T-shaped manner, one portion running into the central nervous system and the other to the periphery of the body. Embryological and comparative anatomical studies have made it seem probable that the peripheral sensory fibre, the process which conducts toward the cell, represents the protoplasmic processes, while that which conducts away from the cell is the axis-

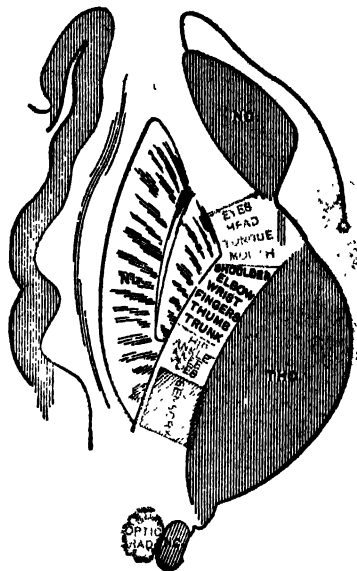


FIG. 18.—DIAGRAM OF MOTOR AND SENSORY REPRESENTATION IN THE INTERNAL CAPSULE. NL., Lenticular nucleus. NC., Caudate nucleus. THO., Optic thalamus. The motor paths are red and black, the sensory are blue.

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cylinder process. In the peripheral sensory nerves we have, then, the dendrites of the lower sensory neurones. These start in the periphery of the body from their various specialized end organs. The axis-cylinder processes leave the ganglia and enter the spinal cord by the dorsal roots of the spinal nerves. After entering the cord each axis-cylinder process divides into an

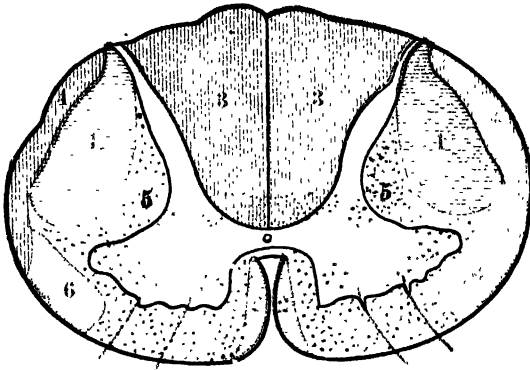


FIG. 19.—DIAGRAM OF CROSS-SECTION OF THE SPINAL CORD, SHOWING MOTOR, RED, AND SENSORY, BLUE, PATHS.

1, Lateral pyramidal tract. 2, Ventral pyramidal tract. 3, Dorsal columns. 4, Direct cerebellar tract. 5, Vento-lateral ground bundles. 6, Vento-lateral ascending tract of Gowers. (Van Gehuchten, colored.)

ascending and a descending branch, which run in the dorsal fasciculi. The descending branch runs but a short distance, and ends in the gray matter of the same side of the cord. It gives off a number of collaterals, which also end in the gray matter. The ascending branch may end in the gray matter soon after entering, or it may run in the dorsal fasciculi as far as the medulla, to end about the nuclei there. In any case it does not cross the middle line. The lower sensory neurone is direct.

The cells about which the axis-cylinder processes and their collaterals of the lower sensory neurone end are of various kinds. They are known as sensory neurones of the second order. In the first place, some of them end about the cell bodies of the lower motor neurones, forming the path for reflexes. They also end about cells whose axis-cylinder processes cross the middle line and run to the opposite side of the brain. In the spinal cord these cells are found in the different parts of the gray matter, and their axis-cylinder processes run in the opposite ventro-lateral ascending tract of Gowers (Fig. 19, 6), and in the ground bundles (fasciculus lateralis proprius and fasciculus ventralis proprius).

In the medulla the nuclei of the dorsal fasciculi (nucleus fasciculi gracilis and nucleus fasciculi cuneati) contain for the most part cells of this character. Their axis cylinder processes, after crossing, run toward the brain in the medial lemniscus or bundle of the fillet; certain of the longitudinal bundles in the formatio reticularis also represent sensory paths from the spinal cord and medulla toward higher centres. The fibres of the medial lemniscus or fillet do not, however, run directly to the cerebral cortex. They end about cells in

## DISEASES OF THE NERVOUS SYSTEM

the ventro-lateral portion of the optic thalamus, and the tract is continued on by way of another set of neurones, which send processes to end in the cortex of the posterior central and parietal convolutions. This is the most direct path of sensory conduction, but by no means the only one. The peripheral sensory neurones may also end about cells in the cord whose axones run but a short distance toward the brain before ending again in the gray matter, and the path, if path it can be called, is made up of a series of these superimposed neurones. The gray matter of the cord itself is also believed to offer paths of sensory conduction. All these paths reach the tegmentum and optic thalamus, and thence are distributed to the cortex along with the other sensory paths. There may also be paths of sensory conduction through the cerebellum by way of the direct cerebellar tract and Gowers' bundle.

From this short summary it is evident that the possible paths for the conduction of afferent impulses are many, and become more complex as the various tracts approach the brain where our knowledge of them is somewhat indefinite. The anatomical arrangement of the two lower orders of sensory neurones is, however, sufficiently well understood to be of great clinical value. In the case of the motor neurones the distribution of the peripheral nerves to the muscles, owing largely to the interlacing into plexuses of the neurones from the various spinal units, is quite different from that of the ventral roots themselves, and the same rule holds true for the peripheral nerve and dorsal root distribution for the cutaneous areas. The *cutaneous* fields corresponding to the peripheral nerves are well known, and although our knowledge of the exact site and outline of some of the *segmental* skin-fields, represented by the dorsal roots, is less accurately established, nevertheless they are sufficiently well understood to be of aid in determining the segmental level of spinal cord and dorsal root lesions. Information concerning the topography in the adult of these skin units or dermatomes has been obtained from various sources: from morphological studies; from anatomical dissections; from physiological experimentation, particularly in Sherrington's hands; from the study of anæsthesias after traumatic injuries to the cord, and from Head's studies of the distribution of the cutaneous lesions in herpes zoster, and of the areas of referred pain and tenderness in visceral disease. The diagrams on pages 928 and 929 embody the results of many of these observations.

The cutaneous sensory impressions are in man conducted toward the brain, probably on the opposite side of the cord—that is, the path crosses to the opposite side soon after entering the cord. Muscular sense, on the other hand, is conducted on the same side of the cord in the fasciculus of Goll to cross above by means of the axones of sensory neurones of the second order in the medulla.

**SENSORY AREAS OF THE BRAIN.**—There are probably two sensory centres—one in the optic thalamus, the other in a considerable area of the cerebral cortex. The thalamus plays a threefold part. Here all the afferent paths terminate; secondly, it contains a mass of gray matter which forms the centre for certain fundamental elements of sensation, particularly those capable of evoking pleasure and discomfort and consciousness of changes of state. Thirdly, in the lateral part of the thalamus is the centre through which the cortex influences the essential thalamic centre, controlling and checking its activity. On their way from the periphery to the cortex afferent impulses

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pay toll to the co-ordinate mechanisms of the spinal cord and the cerebellum. At the thalamic junction they are regrouped to act upon the two terminal centres. One of these, the essential organ of the optic thalamus, responds to all those elements which evoke consciousness of an internal change in state, more particularly pleasure and discomfort. Sensory impulses, then, pass by way of the internal capsule to the cortex, and in the main five groups of sensory impulses are distributed in this way: (1) those underlying postural recognition, and the appreciation of passive movement and weight; (2) the impulses underlying the recognition of tactile differences; (3) those upon which depends the recognition of size and space; (4) those which enable us to localize the spot stimulated; and (5) thermal impulses (Head and Holmes).

These afferent materials are combined in the cortex with each other and with other sense impressions in intellectual processes. The cortical area concerned is that situated between the pre-central fissure and the occipital lobe.

The paths for the conduction of the stimuli which underlie the special senses are given in the section upon the cerebral nerves, and it is only necessary here to refer to what is known of the cortical representation of these senses.

*Visual impressions* are localized in the occipital lobes. The primary visual centre is on the mesial surface in the cuneus, especially about the calcarine fissure, and here are represented the opposite visual half-fields. Some authors believe that there is another higher centre on the outer surface of the occipital lobe, in which the vision of the opposite eye is chiefly represented. However this may be, most authors hold that the angular gyrus of the left hemisphere is a part of the brain in which are stored the memories of the meaning of letters, words, figures, and, indeed, of all seen objects. This is designated as the visual speech centre on the diagram (Fig. 17).

*Auditory impressions* are localized for the most part in the first temporal convolution and the transverse temporal gyri, and it is in this region in the left hemisphere that the memories of the meanings of heard words and sounds are stored. Musical memories are localized somewhat in front of those for words. The cortical centres for smell include a part of the base of the frontal lobe, the uncus, and perhaps the gyrus hippocampi. The centres for taste are supposed to be situated near those for smell, but we possess as yet no definite information about them.

**Topical Diagnosis.**—The diagnosis of the position of a lesion in the nervous system depends upon a careful examination into all the symptoms and signs, and then endeavoring with the help of anatomy and physiology to determine the place, a disturbance at which might produce these symptoms. The abnormalities of motion are usually the most important localizing symptoms, both from the ease with which they can be demonstrated, and also because of the comparative accuracy of our knowledge of the motor path.

Lesions in any part of the motor path cause disturbances of motion. If destructive, the function of the part is abolished, and as the result there is *paralysis*. If the lesion is an irritative one, the structures are thrown into abnormal activity, which produces *abnormal muscular contraction*. The character of the paralysis or of the abnormal muscular contraction varies with lesions of the upper and lower segment, the variations depending, first, upon

# DISEASES OF THE NERVOUS SYSTEM

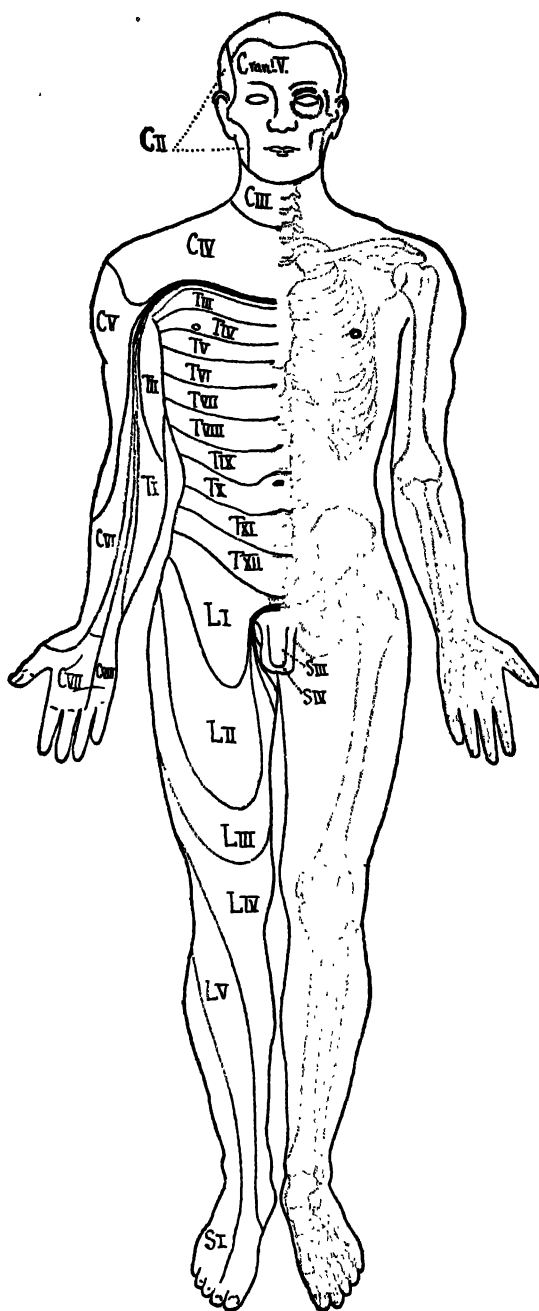


FIG. 20.—ANTERIOR ASPECT OF THE SEGMENTAL SKIN FIELDS OF THE BODY, COMBINED FROM THE STUDIES OF HEAD, KOCHER, STARR, THORBURN, EDINGER, SHERRINGTON, WICHMANN, SEIFFER, BOLK, CUSHING, AND OTHERS.

Heavy lines represent levels of fusion of dermatomes and the pre-axial and postaxial lines of the limbs.

# GENERAL INTRODUCTION

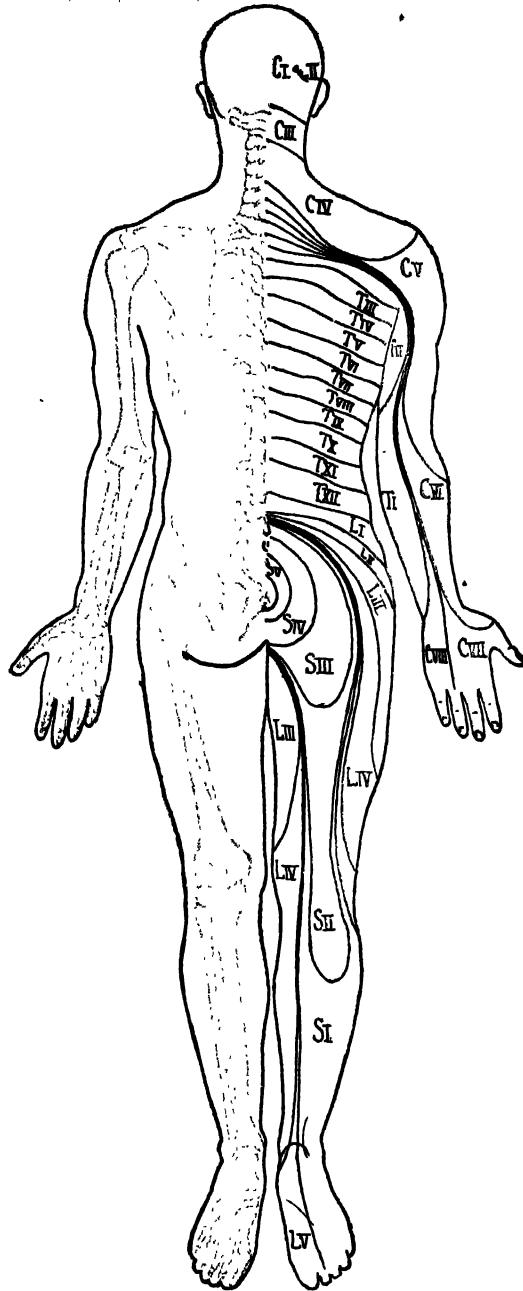


FIG. 21.—POSTERIOR ASPECT OF THE SEGMENTAL SKIN FIELDS OF THE BODY.

the anatomical position of the two segments; and, secondly, upon the symptoms which are the result of secondary degeneration in each of the segments.

(a) LESIONS OF THE LOWER OR SPINO-MUSCULAR SEGMENT.—*Destructive Lesions.*—The nutrition of all parts of a neurone depends upon their connection with its healthy cell body; if the cell body be injured, its processes undergo degeneration, or if a portion of a process be separated from the cell body, that part degenerates along its whole length. This so-called secondary degeneration plays a very important rôle in the symptomatology.

In the lower motor segment the degeneration not only affects the axis-cylinder processes which run in the peripheral nerves, but also the muscle fibres in which the axis-cylinder processes end. The degeneration of the nerves and muscles is made evident, first by the muscles becoming smaller and flabby, and, secondly, by change in their reaction to electric stimulation. The degenerated nerve gives no response to either the galvanic or the faradic current, and the muscle does not respond to faradic stimulation, but reacts in a characteristic manner to the galvanic current. The contraction, instead of being sharp, quick, lightning-like, as in that of a normal muscle, is slow and lazy, and is often produced by a weaker current, and the anode-closing contraction may be greater than the cathode-closing contraction. This is the *reaction of degeneration*, but it is not always present in the classical form. The essential feature is the slow, lazy contraction of the muscle to the galvanic current, and when this is present the muscle is degenerated.

The myotatic irritability, or muscle reflex, and the muscle tonus depend upon the integrity of the reflex arc, of which the lower motor segment is the efferent limb, and in a paralysis due to lesion of this segment the tendon reflexes are abolished and there is a diminished muscular tone.

Lower segment paralyzes have for their characteristics degenerative atrophy with the reaction of degeneration in the affected muscles, loss of their reflex excitability, and a diminished muscular tension. These are the general characteristics, but the anatomical relations of this segment also give certain peculiarities in the distribution of the paralyzes which help to distinguish them from those which follow lesions of the upper segment, and which also aid in determining the site of the lesion in the lower segment itself. The cell bodies of this segment are distributed in groups from the level of the peduncles of the brain throughout the whole extent of the spinal cord to its termination opposite the second lumbar vertebra, and their axis-cylinder processes run in the peripheral nerves to every muscle in the body: so that the component parts are more or less widely separated from each other, and a local lesion causes paralysis of only a few muscles or groups of muscles, and not of a whole section of the body, as is the case where lesions affect the upper segment. The muscles which are paralyzed indicate whether the disease is in the peripheral nerves or spinal cord; for the muscles are represented differently in the peripheral nerves and in the spinal cord. Sensory symptoms, which may accompany the paralysis, are often of great assistance in making a local diagnosis. Thus, in a paralysis with the characteristics of a lesion of the lower motor segment, if the paralyzed muscles are all supplied by one nerve, and the anæsthetic area of the skin is supplied by that nerve, it is evident that the lesion must be in the nerve itself. On the other hand, if the muscles paralyzed are not supplied by a single nerve, but are represented close

together in the spinal cord, and the anæsthetic area corresponds to that section of the cord (see table), it is equally clear that the lesion must be in the cord itself or in its nerve roots.

*Irritative Lesions of the Lower Motor Segment.*—Lesions of this segment cause comparatively few symptoms of irritation. The fibrillary contractions which are so common in muscles undergoing degeneration are probably due to stimulation of the cell bodies in their slow degeneration, as in progressive muscular atrophy, or to irritation of the axis-cylinder processes in the peripheral nerves, as in neuritis. Lesions which affect the motor roots as they leave the central nervous system may cause spasmodic contractions in the muscles supplied by them. Certain convulsive paroxysms, of which laryngismus stridulus is a type and to which the spasms of tetany also belong, are believed to be due to abnormal activity in the lower motor centres. These are the "lowest level fits" of Hughlings Jackson. Certain poisons, as strychnia and that of tetanus, act particularly upon these centres.

The lower motor segment may be involved in all diseases involving the peripheral nerves in cerebral and spinal meningitis, in injuries, in hæmorrhages and tumors of the medulla and cord or their membranes, in lesions of the gray matter of the segment, in anterior poliomyelitis, progressive muscular atrophy, bulbar paralysis, ophthalmoplegia, syringomyelia, etc.

(b) LESIONS OF THE UPPER MOTOR SEGMENT.—*Destructive lesions* cause paralysis, as in the lower motor segment, and here again the secondary degeneration which follows the lesion gives to the paralysis its distinctive characteristics. In this case the paralysis is accompanied by a *spastic* condition, shown in an exaggeration of muscle reflex and an increase in the tension of the muscle. It is not accurately known how the degeneration of the pyramidal fibres causes this excess of the muscle reflex. The usual explanation is that under normal circumstances the upper motor centres are constantly exerting a restraining influence upon the activity of the lower centres, and that when the influence ceases to act, on account of disease of the pyramidal fibres, the lower centres take on increased activity, which is made manifest by an exaggeration of the muscle reflex.

The neurones composing each segment of the motor path are to be considered as nutritional units, and therefore the secondary degeneration in the upper segment stops at the beginning of the lower. For this reason the muscles paralyzed from lesions in the upper segment do not undergo degenerative atrophy, nor do they show any marked change in their electrical reactions.

The separate parts of the upper motor segment lie much more closely together than do those of the lower segment, and therefore a small lesion may cause paralysis in many muscles. This is more particularly true in the internal capsule, where all the axis-cylinder processes of this segment are collected into a compact bundle—the pyramidal tract. A lesion in this region usually causes paralysis of most of the muscles on the opposite side of the body—that is, hemiplegia. The pyramidal tract continues in a compact bundle, giving off fibres to the motor nuclei at different levels; a lesion anywhere in its course is followed by paralysis of all the muscles whose spinal centres are situated below the lesion. When the disease is above the decussation, the paralysis is on the opposite side of the body; when below, the paralyzed muscles are on the same side as the lesion. Above the internal capsule the path is

somewhat more separated, and in the cortex the centres for the movements of the different sections of the body are comparatively far apart, and a sharply localized lesion in this region may cause a more limited paralysis, affecting a limb or a segment of a limb—the cerebral monoplegias; but even here the paralysis is not confined to an individual muscle or group of muscles, as is commonly the case in lower segment paralysis (see Fig. 15 and explanation).

To sum up, the paralyzes due to lesions of the upper motor segment are widespread, often hemiplegic; the paralyzed muscles are spastic (the tendon reflexes exaggerated), they do not undergo degenerative atrophy, and they do not present the degenerative reaction to electrical stimulation.

*Irritative Lesions of the Upper Motor Segment.*—Our knowledge of such lesions is confined for the most part to those acting on the motor cortex. The abnormal muscular contractions resulting from lesions so situated have as their type the localized convulsive seizures classed under *Jacksonian* or cortical epilepsy, which are characterized by the convulsion beginning in a single muscle or group of muscles and involving other muscles in a definite order, depending upon the position of their representation in the cortex. For instance, such a convulsion, beginning in the muscles of the face, next involves those of the arm and hand, and then the leg. The convulsion is usually accompanied by sensory phenomena and followed by a weakness of the muscles involved.

A majority of lesions of the motor cortex are both destructive and irritative—i. e., they destroy the nerve cells of a certain centre, and either in their growth or by their presence throw into abnormal activity those of the surrounding centres.

The upper motor segment is involved in nearly all the diseases of the brain and spinal cord, especially in injuries, tumors, abscesses, and hæmorrhages; transverse lesions of the cord; syringomyelia, progressive muscular atrophy, bulbar paralysis, etc. One lesion often involves both the upper and the lower motor segments, and there is paralysis in the different parts of the body, with the characteristics of each. Such a combination enables us in many cases to make an accurate local diagnosis.

Lesions in the optic path and in the different speech centres also give localizing symptoms, which should always be looked for.

(c) LESIONS OF THE SENSORY PATH.—Here again the lesion may be either irritative or destructive. *Irritative lesions* cause abnormal subjective sensory impression—paræsthesia, formication, a sense of cold or constriction, and pain of every grade of intensity. The character of the sensory symptoms gives very little indication as to the position of the irritating process. Intense pain is, as a rule, a symptom of a lesion in the peripheral sensory neurones, but it may be caused by a disease of the sensory path within the central nervous system.

The exact distribution of symptoms gives more accurate data, for if they are confined to the distribution of a peripheral nerve or of a spinal segment the indication is plain. If one side of the body is more or less completely affected, the lesion is somewhere within the brain, etc.

*Destructive Lesions.*—A complete destruction of the sensory paths from any part of the body would of course deprive that part of sensation in all its qualities. This occurs most frequently from injury to the peripheral sensory neurones within the peripheral nerves, and the area of anæsthesia depends

upon the nerve injured. Complete transverse lesion of the cord causes complete anæsthesia below the injury.

Unilateral lesions of the cord, medulla, dorsal part of the pons, tegmentum, thalamus, internal capsule, and cortex cause disturbances of sensation on the *opposite* side of the body; here again the extent of the defect more than its character helps us to determine the position of the lesion. Hemianæsthesia involving the face as well as the rest of the body can only occur above the place where the sensory paths from the fifth nerve have crossed the middle line on their way to the cortex. This is in the upper part of the pons. From this point to where they leave the internal capsule the sensory paths are in fairly close relation, and are at times involved in a very small lesion. Above the internal capsule the paths diverge quickly, and for this reason only an extensive lesion can involve them all, and in lesions of this part we are more apt to have the sensory disturbances confined to one or another region of the body. Unilateral lesions of the thalamus, pons, medulla, and cord usually cause sensory disturbances on the same side of the body, as well as those on the opposite side. These are due to the involvement of the sensory paths as they enter the central nervous system at or a little below the site of the lesion and before the axones of the sensory neurones of the second order have crossed the middle line. The area of disturbed sensation on the same side is limited to the distribution of one or more spinal segments and often indicates accurately the position and extent of the diseased process. As a rule, destructive lesions of the central nervous system do not involve all the paths of sensory conduction, and the loss of sensation is not complete. It is often astonishing how very slight the sensory disturbances are which result from an extensive lesion. Sensation may be diminished in all of its qualities, or, what is more common, certain qualities may be affected while others are normal. Thus, the sense of pain and temperature may be lost while that of touch remains normal, as is often the case in diseases of the spinal cord, or there may be simply a loss of the muscular sense and of the stereognostic sense (the complex sensory impression which enables one to recognize an object placed in the hand), as occurs frequently from lesions of the cortex. Occasionally pain sensation persists with loss of tactile and thermic sensations. Almost every other combination has been described. It is the *distribution* more than the character of the sensory defect that is of importance, and often the distribution gives but uncertain indication of the position of the lesion. The combination of the sensory defect with different forms of paralysis gives the most certain diagnostic signs.

*Sympathetic Nervous System (Involuntary, Vegetative, Visceral, Autonomic).*—This system innervates the pupils, non-striated muscles, glands, viscera, heart and blood vessels, and genital organs. It is outside the control of the will but can be influenced by the central nervous system, especially by emotional stimuli. The reverse may occur; disturbance in the realm of the sympathetic system may affect the general nervous system. Digestive disturbance may result from nervous anxiety or fatigue, and conversely disturbed function of the alimentary tract may cause marked nervous depression.

This involuntary or vegetative nervous system consists of two parts which are distinct anatomically and antagonistic physiologically.

## DISEASES OF THE NERVOUS SYSTEM

(1) Sympathetic proper (thoracico-lumbar).

(2) Para-sympathetic (a) cranio-bulbar and (b) sacral.

There is some confusion in the use of the term "autonomic" which was applied by Langley to the whole vegetative system, but is also used by some to designate the para-sympathetic alone.

The fibres of the sympathetic proper arise from cells in the intermediolateral region of the cord (preganglionic), pass by the anterior roots to end in ganglia which in turn send fibres (postganglionic) to the terminations in smooth muscle, the heart, blood-vessels, sweat glands, secreting glands, etc. The receptor (afferent) elements are concerned with visceral sensations and referred visceral pain. The excitor (efferent) elements form synapses in the ganglia and in this way one fibre may stimulate a number of cells. From these cells the postganglionic fibres pass directly to their destinations. The ganglia act as "distributing stations" and form a series in front of the vertebral column, one on each side. In the neck there are three ganglia in each chain, connected with the cord through the first and second thoracic roots. In the thoracic, lumbar and sacral regions there is a ganglion for each nerve root.

The para-sympathetic system (often termed autonomic or system of the "extended vagus") has the ganglia placed more peripherally. In the *cranio-bulbar* portion, fibres pass from the mid-brain to the ciliary ganglion, constricting the pupil, from the medulla secretory fibres go to the submaxillary glands and by the vagus inhibitory fibres go to the heart, constrictor to the bronchi, motor to the oesophagus, stomach and intestines, and secretory to the stomach and intestines. The vague nerve is the most important constituent of the para-sympathetic system. From the sacral portion by the pelvic nerve fibres go to the descending colon, rectum, anus, bladder and genital system. The vegetative system has three plexuses, cardiac, solar and hypogastric, which receive fibres from both systems. The sympathetic system has close relations to the endocrine glands. (1) The thyroid, adrenals and pituitary glands are in close relationship to the thoracico-lumbar sympathetic and accelerate metabolism (*katabolic*). (2) The para-sympathetic has close relations to the digestive tract and its glands and is engaged in storing energy (*anabolic*).

When the sympathetic and para-sympathetic supply the same structure, their influences are antagonistic. Thus the sympathetic dilates the pupil, the other contracts it; the sympathetic increases the heart rate, the other slows it; the sympathetic inhibits the movements of the gastro-intestinal tract, the other increases them. In conditions of health there is a balance between the two systems. To describe the resulting condition when this balance is disturbed the terms sympathicotonia and vagotonia are employed, depending on which system is over-active. In the diagnosis of this the effects of certain drugs are important. Thus the sympathetic system proper is stimulated by epinephrine (1 c. c. of 1-1000 solution) with resulting tremor, rigor, a sense of cold, glycosuria and a rise in blood pressure. The para-sympathetic system is stimulated by pilocarpine (gr. 1/20-1/6, 0.003-0.01 gm.) with resulting salivation, nausea, sweating, flushing and a fall in blood pressure. Atropine (gr. 1/100-1/50, 0.00065-0.0013 gm.) paralyses the para-sympathetic system with resulting dryness of the mouth and throat, palpitation and oppression.

Clinically among the features of vagotonia are small pupils, salivation,

flushing, sweating, clammy hands and feet, dermatographia, bradycardia, irregularity of respiration, hyperacidity, cardio- and pylorospasm, spastic constipation, and sphincter contraction. There may be increased sensitiveness to foreign proteins, as shown by urticaria, anaphylaxis; some include asthma. Among those of sympathicotonia are, dilated pupils, prominence of the eyes, dryness of the mouth and dry skin, tachycardia, decreased sugar tolerance and atony of the digestive tract. Actually it is found that many patients show features suggestive of disturbance in both systems. Some show vagotonia at one time and sympathicotonia later. The sympathetic system stands in close relation to the endocrine glands and its stimulation may cause increased activity of the adrenal and thyroid glands particularly.

## B. SYSTEM DISEASES

### I. INTRODUCTION

There are certain diseases of the nervous system which are confined, if not absolutely, still in great part, to definite tracts (combinations of neurones) which subserve like functions. These tracts are called *systems*, and a disease which is confined to one of them is a *system disease*. If more than one system is involved, the process is called a combined system disease. Just what diseases should be classed under these names has given rise to much discussion but little agreement. We can not speak positively; our knowledge is not sufficiently accurate, either as to the exact limits of the systems themselves, or the nature and extent of the process in the several diseases.

It may be said that the nervous system is composed of two great systems of neurones, the afferent or sensory system and the efferent or motor system, and the connections between them. (See General Introduction.)

Tabes dorsalis is a disease confined at its onset to the afferent system, and progressive muscular atrophy is one of the efferent system. Several theories have been advanced to explain why a disease should be limited to a definite system of neurones. One view is based upon the idea that in certain individuals one or the other of these systems has an innate tendency to undergo degeneration; another assumes that neurones with a similar function have a similar chemical construction (which differs from that of neurones with a different function) and this is taken to explain why a toxin should show a selective action for a single functional system of neurones.

## II. DISEASES OF THE AFFERENT OR SENSORY SYSTEM

### I. TABES DORSALIS

(*Locomotor Ataxia; Posterior Spinal Sclerosis*)

**Definition.**—An affection characterized clinically by sensory disturbances, incoördination, trophic changes, and involvement of the special senses, par-

ticularly the eyes. Anatomically there are found degenerations of the root fibres of the dorsal columns of the cord, of the dorsal roots, and at times of the spinal ganglia and peripheral nerves. Degenerations have been described in the brain, particularly the cortex cerebri, in the ganglion cells of the cord, and in the endogenous fibres of the dorsal columns.

**Etiology.**—It is a widespread disease, more frequent in cities than in the country. Among 16,562 cases in the neurological dispensary of the Johns Hopkins Hospital there were 201 cases of tabes. Males are attacked more frequently than females, the proportion being nearly 10 to 1. The disease is not very uncommon in the negro in the United States. It is a disease of adult life, the great majority of cases occurring between the thirtieth and fiftieth years. There are a good many cases of the existence of the disease in both husband and wife, and a few in which the children were also affected. Occasionally cases are seen in young men, and it may occur in children with congenital syphilis. *Syphilis* is the important cause. There is evidence which suggests that certain strains of the *Treponema* are particularly likely to attack the nervous system. The interval between the syphilitic infection and the first symptoms of tabes is variable. Five to fifteen years is the period in one-half the cases. Intervals from two to twenty-five years occur.

**Pathology.**—Posterior spinal sclerosis, although the most obvious gross change, is not an adequate description. The dorsal fibres are of two kinds, those with their cell bodies outside the cord in the spinal ganglia, the so-called exogenous, or root fibres, and those which arise from cells within the cord, the endogenous fibres. These two sets occupy fairly well-determined regions, and a study of early cases of tabes has shown that it is the exogenous or root fibres that are first affected. The fibres of the dorsal roots enter the cord in two divisions, an external and an internal; the former is composed of fibres of small calibre, which, in the cord, make up Lissauer's tract, and occupy the space between the apex of the dorsal cornua and the periphery of the cord, and really do not form part of the dorsal columns. They are short, soon entering the gray matter, and do not seem to be affected, or only slightly so, in early cases.

The larger fibres enter the cord by the internal division, just medial to the cornua, in what is known as the root entry zone. Some enter the gray matter of the spinal cord almost directly and others after a longer course, while still others run in the cord to the medulla, to end in the nuclei of the dorsal columns. As the fibres of every spinal nerve enter the cord between the dorsal cornua and the nerve fibres which have entered lower down, the fibres from each root are successively pushed more and more toward the median line, and so in the cervical cord the fasciculi of Goll are largely composed of long fibres derived from the sacral and lumbar roots.

That it is the coarse dorsal root fibres which are first affected in tabes is generally admitted, but there is much divergence of opinion as to the character and location of the initial process.

Nageotte calls attention to the frequency of a transverse, interstitial neuritis of the dorsal roots just after they have left the ganglia and are still surrounded by the dura, and he believes that it is this neuritis which is the primary lesion. Oppenheimer and Redlich laid stress on the presence of inflammation of the pia mater over the dorsal aspect of the cord, which involves the

root fibres as they pass through. They point out that it is just here that the dorsal roots are most vulnerable, for at this point—that is, while surrounded by the pia—they are almost completely devoid of their myelin sheaths. Changes in the blood-vessels of the cord, of the pia, and of the nerve roots have been described in early tabes, and Marie and Guillain advanced the belief that the changes in the cord are due to syphilis of the posterior lymphatic system which is confined to the dorsal columns of the cord, the pia mater over them, and the dorsal roots. For them the changes in the nervous system are only apparently radicular or systemic.

With the Marchi stain, degeneration of the root fibres in the root-entry zone is a constant finding in early cases. This change is radicular in the sense that it varies in intensity with the different roots and is most marked in the sacral and lumbar regions. The degeneration is not found in the dorsal roots, but begins within the cord just beyond where the root fibres lose their neurilemma and their myelin sheaths. Degenerated fibres may be traced into the dorsal gray matter and among the ganglion cells of the columns of Clarke. The long columns which ascend the cord also degenerate.

In more advanced cases, there are degeneration of the dorsal roots and some alteration of the cells in the spinal ganglia. The fibres distal to the ganglia are practically normal, although at times the sensory fibres, at the periphery of a limb, show degeneration. Within the cord, the exogenous fibres are diseased; there is also degeneration in the endogenous system of fibres. Optic atrophy is frequently found. The other cranial nerves, especially the fifth with its ganglion, have been found degenerated.

The disease occasionally spreads beyond the sensory system in the cord, and in advanced cases the cells in the ventral horns may be degenerated in association with muscular atrophy. Mott very generally found more or less marked changes in the pyramidal fibres; these he believed to be evidence of changes in the cerebral cortex. Degeneration of the cortex may exist, but even when mental symptoms are absent, or very mild, similar slight changes have been described, just as in general paresis, without marked tabetic symptoms, there may be degeneration of the dorsal columns. The close association of tabes and general paresis will be considered later.

**Symptoms.**—For convenience, these are considered under three stages—the incipient or pre-ataxic, the ataxic and the paralytic.

**INCIDENT STAGE.**—The onset differs very widely in the different cases, and mistakes in diagnosis are often made early in the disease. The following are the most characteristic initial symptoms:

*Pains*, usually of a sharp stabbing character; hence, the term lightning pains. They last for only a second or two and are most common in the legs or about the trunk, and tend to follow dorsal root areas. They dart from place to place. At times they are associated with a hot burning feeling and often leave the affected area painful to pressure, and occasionally herpes may follow. The intensity of the pain varies from a sore, burning feeling of the skin to a pain so intense that, were it not for momentary duration, it would exceed human endurance. They occur at irregular intervals, and are prone to follow excesses or to come on when health is impaired. When typical, these pains are practically pathognomonic. Gastric and other crises may occur. Paræsthesia may be among the first symptoms—numbness of the feet, tin-

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ting, etc.—and at times a sense of constriction about the body. Hyperaesthesia is common on the trunk, especially in the lower part, and with it the abdominal reflexes may be very active. There may be areas over which there is marked tenderness on any stimulus, especially cold. There may be subjective loss of sensation in various areas.

*Objective sensory* changes appear early and are always present if subjective pain has been present for any time. The areas may be very sensitive to stimuli and with this there may be decreased pain sensibility. The disturbances are variable but pain and temperature sensations are often affected.

*Ocular Symptoms.*—(a) *Optic atrophy.* This occurs in about 10 per cent. of the cases, and is often an early and even the first symptom. There is a gradual loss of vision, which in a majority of cases leads to total blindness. This appears to be secondary to a syphilitic meningitis. (b) *Ptosis*, which may be double or single. (c) *Paralysis* of the external muscles of the eye. This may be of one muscle or occasionally of all the muscles of the eye. The paralysis is often transient, the patient merely complaining that he saw double for a certain period. (d) *Argyll-Robertson pupil*, in which there is loss of the iris reflex to light but contraction during accommodation. The pupils are often very small—spinal myosis—and may be unequal or irregular.

*Bladder Symptoms.*—The first warning may be difficulty in emptying the bladder. Incontinence of urine occurs only at a later stage. Decrease in sexual desire and power may be an early symptom.

*Trophic Disturbances.*—These usually occur later, but at times they are early symptoms, and one's attention may be called to the trouble by a perforating ulcer or a characteristic Charcot's joint.

*Loss of the Deep Reflexes.*—This early and important sign may occur years before the development of ataxia. Even alone it is of great moment, since it is very rare to meet with individuals in whom the knee and ankle jerks are normally absent. The combination of loss of either of these with one or more of the features mentioned above, especially the lightning pains and ptosis or Argyll-Robertson pupil, is practically diagnostic. These reflexes gradually decrease, and one may be lost before the other, or disappear first in one leg.

These initial symptoms may persist for years without the development of incoördination. The patient may look well and feel well, and be troubled only by the lightning pains or one of the other subjective symptoms. Progressive nerve deafness and paralysis of the vocal cords, with the laryngeal muscles paralyzed or parietic, may occur. The disease may never progress beyond this stage, and when optic atrophy develops early and leads to blindness, ataxia rarely, if ever, supervenes, an antagonism noted by many authors.

*ATAXIC STAGE.*—*Motor Symptoms.*—The ataxia, which comes on gradually, is believed to be due to a disturbance or loss of the afferent impulses from the muscles, joints, and deep tissues. A disturbance of the muscle sense itself can usually be demonstrated. One of the first indications is inability to get about readily in the dark or to maintain equilibrium when washing the face with the eyes shut. When the patient stands with the feet together and the eyes closed, he sways and has difficulty in maintaining his position (Rom-

berg's symptom), and he may be quite unable to stand on one leg. He does not start off promptly at the word of command. On turning quickly he is apt to fall. He has more difficulty in descending than ascending stairs. Gradually the characteristic ataxic gait develops. The normal man walks by faith, the tabetic by sight. The patient, as a rule, walks with a stick, the eyes are directed to the ground, the body is thrown forward, and the legs are wide apart. In walking, the leg is thrown out violently, the foot is raised too high and is brought down in a stamping manner with the heel first, or the whole sole comes in contact with the ground. Ultimately the patient may be unable to walk without the assistance of two canes. This gait is very characteristic, and unlike that seen in any other disease. The inco-ordination is not only in walking, but in the performance of other movements. If the patient is asked, when in the recumbent posture, to touch one knee with the other foot, the irregularity of the movement is very violent. Inco-ordination of the arms is less common, but usually develops in some grade. It may in rare instances exist before the inco-ordination of the legs. It may be tested by asking the patient to close his eyes and to touch the tip of the nose or the tip of the ear with the finger, or with the arms thrust out to bring the tips of the fingers together. The inco-ordination may be noticed early by a difficulty in buttoning the collar or performing one of the routine acts of dressing.

With marked inco-ordination there is but little loss of muscular power. The grip of the hands may be strong and firm, the power of the legs may be unimpaired, and their nutrition, except toward the close, may be unaffected.

There is a remarkable muscular relaxation (*hypotonia*) which enables the joints to be placed in positions of hyperextension and hyperflexion. It gives sometimes a marked backward curve to the legs.

*Sensory Symptoms.*—The lightning pains may persist. They vary greatly in different cases. Some patients are rendered miserable by their frequent occurrence; others escape altogether. In addition, common symptoms are tingling, pins and needles, particularly in the feet, and areas of hyperæsthesia or anæsthesia. The patient may complain of a change in sensation in the soles of the feet, as if cotton was interposed between the floor and the skin. Sensory disturbances occur less frequently in the hands. Objective sensory disturbances can usually be demonstrated, and almost every variety of disturbance of tactile, pain and temperature sense has been described. Bands of a moderate grade of anæsthesia about the trunk are not uncommon; they are apt to follow the distribution of spinal segments. The most marked disturbances are usually found on the legs. Retardation of the sense of pain is common, and a pin prick on the foot is first felt as a simple tactile impression, and the sense of pain is not perceived for a second or two or may be delayed for as much as ten seconds. The pain felt may persist. A curious phenomenon is the loss of the power of localizing pain. If the patient is pricked on one limb he may say that he feels it on the other (*allocheiria*), or a pin prick on one foot may be felt on both feet. Pruritus may occur over the areas affected by the pains. The muscular sense, usually affected early, becomes much impaired and the patient no longer recognizes the position in which his limbs are placed. This may be present in the pre-ataxic stage.

*Reflexes.*—The loss of the knee and ankle-jerks, the latter usually first, is an important sign but occasionally they are retained, and in these cases

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the lumbar segments are little if at all involved. The skin reflexes are often increased early but may be diminished later when the tactile sense is lost. The plantar reflex is usually normal unless there is sclerosis of the pyramidal tracts. The oculo-cardiac reflex is often absent.

**Special Senses.**—The eye symptoms noted above may be present, but ataxia is rare with optic atrophy. Deafness may occur, due to lesion of the auditory nerve. There may also be attacks of vertigo. Olfactory symptoms are rare. W. B. Swift has drawn attention to a voice sign which consists in ataxic speech with "a slovenly indistinct enunciation that shows partially in the vowels but predominantly in the consonants." Suggested tests are "e" (as in ell), "t," "journals" and "Time and tide wait for no man."

**Visceral Symptoms.**—Among the most remarkable sensory disturbances are the *tabetic crises*, severe paroxysms of pain referred to various viscera; thus, ocular, laryngeal, gastric, nephritic, rectal, urethral, and clitoral crises have been described. The most common are the gastric and laryngeal. *Gastric crises* may occur early and persist as the most prominent feature. Starr found them as the first symptom 18 times in 450 cases. The onset is usually sudden, with severe pain in the epigastrium, radiating to the back and behind the sternum. Vomiting follows and may be quite independent of food. Hæmatemesis may occur, not necessarily due to ulcer. Pallor, sweating, cold extremities, and a small pulse are associated, and in rare instances death occurs in collapse. The blood pressure may be very high, as reported by Barker, and it may be that the condition is associated with angiospasm in the gastric and mesenteric vessels. The X-ray examination shows spasmodic contractions of the stomach. No special change may be found at autopsy. In the laryngeal crises there may be true spasm with dyspnoea and noisy inspiration. A patient may die in the attack. There are also nasal crises, associated with sneezing fits. The contrary condition may occur, that is absence of pain from visceral lesions, as rupture of a gastric ulcer, and render diagnosis very difficult.

The *sphincters* are frequently involved. Early in the disease there may be a retardation or hesitancy in making water. Later there is retention, and cystitis may occur. Unless great care is taken the inflammation may extend to the kidneys. Constipation is extremely common. Later the sphincter anal is weakened. The sexual power is usually lost in the ataxic stage.

**Trophic Changes.**—Herpes, œdema, ecchymosis, or local sweating may occur in the course of the lightning pains. Alteration in the nails may occur. A perforating ulcer may develop on the foot, usually beneath the great toe. A perforating buccal ulcer has also been described. Onychia may prove very troublesome.

**Arthropathies (Charcot's Joints).**—Anatomically there are: (1) enlargement of the capsule with thickening of the synovial membranes and effusion; (2) slight enlargement of the ends of the bones, with slight exostoses; (3) a dull velvety appearance of the cartilages, with atrophy in places. The knees are most frequently involved. The spine is affected in rare instances usually in the lumbar region. Trauma, with loss of pain sense, is an important element in the causation. A striking feature is the usual absence of pain. Occasionally there is pain from the distended soft parts and skin but not from the bones. Suppuration may occur, also spontaneous fractures.

*Atrophy* of the muscles, usually a late manifestation, may be localized and associated with neuritis or due to involvement of the ventral horns.

*Aneurism* is found in as high as 20 per cent. of some series, and aortic insufficiency is common. Both are associated syphilitic manifestations.

*Cerebral Symptoms.*—Hemiplegia may develop at any stage of the disease, more commonly when it is well advanced. It may be due to hæmorrhagic softening from disease of the vessels, to progressive cortical changes or rarely to coarse syphilitic disease. The lost knee-jerk may return on the affected side. *Hemianæsthesia* is sometimes present.

*Cerebro-Spinal Fluid.*—The examination is of great value; the findings are: (1) *Cell content.* Lymphocytosis is found in about 90 per cent., the number of cells usually being between 40 and 60, and rarely over 100. The higher counts are found when irritative symptoms are marked. With an arrest of the process the counts are lower. (2) *Globulin.* This is positive in 90-95 per cent. In old quiescent cases there may be no increase. (3) *Wassermann reaction.* This is nearly always positive but may be negative in quiescent cases. The blood Wassermann test is positive in about 70 per cent. (4) *Colloidal gold reaction.* This is present in 85-90 per cent. and is useful in diagnosing tabes from paresis. A paretic curve in a patient with signs of tabes points to the possible development of paresis subsequently.

*PARALYTIC STAGE.*—After persisting for an indefinite number of years the patient gradually becomes bedridden and paralyzed. In this condition he is likely to be carried off by some intercurrent affection, such as pyelo-nephritis, pneumonia, or tuberculosis.

*Juvenile tabes* is more frequent in girls and usually due to congenital syphilis. Optic atrophy is comparatively common, while pains, ataxia and visceral lesions are less frequent than in adults.

*COURSE.*—A patient may remain in the pre-ataxic stage for an indefinite period; and the loss of knee-jerks and atrophy of the optic nerves may be the sole indications of the disease. In such cases inco-ordination rarely develops. In a majority of cases the progress is slow, and after six or eight years, sometimes less, the ataxia is well marked. The symptoms vary a good deal; thus, the pains, which may have been excessive at first, often lessen. The disease may remain stationary for years; then exacerbations occur and it makes rapid progress. Occasionally the process seems to be arrested. There are instances of what may be called acute ataxia, in which, within a year or even less, the inco-ordination is marked, and the paralytic stage may develop within a few months. The disease itself rarely causes death, and after becoming bedridden the patient may live for fifteen or twenty years.

*Diagnosis.*—In well marked cases there is no difficulty but one should not wait for the loss of reflexes or pupil signs to make the diagnosis. The pains are suggestive, especially with a history or evidence of syphilis. The greatest importance in early diagnosis attaches to disturbances of sensation, for which a careful search should be made. The most useful test is the response to the prick of a pin, shown by delay in perception, decreased sensation, or unusual sensations. Certain areas are especially important; the earliest loss is usually on the legs, most marked distally, decreasing toward the trunk and usually not corresponding to root distribution. There is often a zone of analgesia around the trunk, most often on the upper thorax. The

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ulnar border of the arms may be affected or the centre of the face, including the nose and adjoining parts of the cheeks. This may involve the greater part of the face—the tabetic mask. Absence of pain when the calf muscles are compressed and hypotonia are aids. Reduction in the time of appreciation of vibrations of a fixed amplitude over the lower part of the body, especially over the sacrum, is of value. The early ocular palsies are of great importance. A squint, ptosis, or the Argyll-Robertson pupil may be the first sign, and exist with the loss only of the ankle or knee-jerk. Loss of the knee-jerk does occasionally occur in healthy individuals. The Wassermann reaction and a study of the spinal fluid are of great aid.

The diseases most likely to be confounded with tabes dorsalis are: (a) PERIPHERAL NEURITIS.—The steppage gait of arsenical, alcoholic, or diabetic paralysis is quite unlike that of tabes. There is a paralysis of the feet, and the leg is lifted high so that the toes may clear the floor. The use of the word *ataxia* in this connection should not be continued. In the rare cases in which the muscle sense is particularly affected and in which there is true ataxia, the absence of the lightning pains and eye signs, with the history make the diagnosis clear. In diphtheritic paralysis the early loss of the knee-jerk and the eye signs may suggest tabes, but the history, the existence of paralysis of the throat, and the absence of pains render a diagnosis easy.

(b) COMBINED SCLEROSIS.—Marked inco-ordination with spastic paralysis occurs in this condition. In a majority of the cases this is distinguished by the absence of pains and eye signs, but it may be a manifestation of the cord lesions in tabo-paralysis.

(c) CEREBRAL DISEASE.—In diseases involving the afferent tracts ataxia is prominent at times. It is usually unilateral or limited to one limb; this, with the history and the associated symptoms, excludes tabes.

(d) CEREBELLAR DISEASE.—The cerebellar inco-ordination has only a superficial resemblance to that of tabes, and is more a disturbance of equilibrium than a true ataxia; the knee-jerk is usually present, there are no lightning pains or sensory disturbances; while, on the other hand, there are headache, optic neuritis, and vomiting.

(e) SYPHILITIC AFFECTIONS involving the dorsal columns of the cord may be associated with inco-ordination and resemble tabes very closely.

(f) GENERAL PARESIS.—Though of identical origin and often associated, it is of great practical importance to determine, if possible, whether the type is to be spinal or cerebral, for when this is established, it does not often change. The difficulty arises in the premonitory stage, when any alteration in the mental characteristics is of the utmost significance. Loss of the deep reflexes and lightning pains speak for tabes; active reflexes, with ocular changes, especially optic atrophy, are suggestive of paresis.

(g) VISCERAL CRISES and NEURALGIC SYMPTOMS may lead to error, and in middle-aged men with severe, recurring attacks of abdominal pain, it is always well to bear in mind the possibility of tabes.

**Prognosis.**—Complete recovery can not be expected, but arrest of the process is not uncommon and a marked amelioration is frequent. Optic-nerve atrophy, one of the most serious events in the disease, has this hopeful aspect—that inco-ordination rarely follows and the progress of the spinal symptoms may be arrested. On the other hand, mental symptoms are more likely to

follow. The optic atrophy itself is occasionally checked. On the whole, the prognosis is bad. There is more hope that in early cases coming on soon after infection the course may be arrested. Death is usually from some cardiovascular complication; next in frequency from tuberculosis and pneumonia.

**Treatment.**—To arrest the progress and to relieve, if possible, the symptoms are the objects which the practitioner should have in view. A quiet, well-regulated method of life is essential. It is not well, as a rule, for a patient to give up his occupation so long as he is able to keep about and perform ordinary work, provided there is no evident mental change. Tabetics have for years conducted large businesses, and there have been several notable instances in our profession of men who have risen to distinction in spite of the existence of this disease. Care should be taken in the diet, particularly if gastric crises have occurred. Attention should be given to the bladder and bowels. Excesses of all sorts, more particularly in *baccho et venere*, should be carefully avoided. A man in the pre-ataxic stage should not marry.

To secure arrest of the disease many remedies have been employed.

As soon as tabes is diagnosed or strongly suspected active treatment should be given; mercury is probably of more value than arsphenamine. A thorough course of mercurial inunctions should be given (20 to 30) and neo-arsphenamine given intravenously (0.3-0.6 gm.) once a week for six weeks. This sequence should be repeated once or twice and its further repetition decided by the condition. Iodides should be given steadily in full dosage. The treatment should be carried out persistently and the spinal fluid studied frequently as improvement in this is a good indication. If there is no change or only slight gain the intraspinal treatment with auto-arsphenaminized serum may be used but the enthusiasm for this is less. In some cases the giving of mercury by inunction followed by spinal puncture weekly, in which as much serum is withdrawn as possible has proved of benefit. The use of mercurial serum has been helpful in some cases.

For the pains, complete rest in bed and counter-irritation to the spine may be employed. The severe spells which come on particularly after excesses of any kind are often relieved by a hot bath or by a Turkish bath. For the severe recurring attacks of lightning pains spinal cocainization may be tried. Cannabis indica, acetyl-salicylic acid, phenacetine, etc., are sometimes useful. Suppositories of codeine (gr. 1, 0.06 gm.) and extract of belladonna (gr. 1/2, 0.03 gm.) may give relief. In the severe paroxysms of pain and the crises morphia may be necessary but its use should be as infrequent as possible. Electricity is of very little benefit. For gastric crises, chloretone (gr. v-x, 0.3-0.6 gm.) or tincture of iodine (℥ x, 0.6 c. c.) may be given. The dorsal spinal nerve roots of the seventh, eighth, ninth, and tenth have been divided with good results. The laryngeal crises are rarely dangerous. An application of cocaine may be made or a few whiffs of chloroform or nitrite of amyl may be given. In all cases of tabes with hypertension the prolonged use of nitroglycerine, given until the physiological effect is produced, is of great service in allaying pain and diminishing the frequency of the crises. Its use must be guarded when there is aortic insufficiency. The bladder symptoms demand constant care. When the organ can not be perfectly emptied the catheter should be used, and the patient may

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be taught its use and how to keep it thoroughly sterilized. Tincture of belladonna (m v-x, 0.3-0.6 c. c.) is useful when there is incontinence.

Frenkel's method of *re-education* often helps the patient to regain to a considerable extent the control of the voluntary movements. The patient is first taught, by repeated systematic efforts, to perform simple movements; from this he goes to more and more complex movements. The treatment should be directed and supervised by a trained teacher, as the result depends upon the skill of the teacher quite as much as upon the perseverance of the patient.

### II. GENERAL PARESIS AND TABO-PARALYSIS

The majority of cases of tabes run their course with practically no mental symptoms, and cases of general paresis may never present symptoms that suggest tabes. For practical purposes we are forced to keep the distinction clearly in mind, and it seems best to consider them separately. There is, however, a group of cases in which the symptoms of the two diseases are associated in every combination for which the name "tabo-paralysis" is used.

#### *General Paresis*

**Definition.**—A chronic meningo-encephalitis caused by the spirochæte of syphilis, often associated with other local changes leading to mental disturbances and finally to dementia and paralysis.

**Etiology.**—The average interval from the syphilitic infection is twelve years. Males are affected much more frequently than females. It occurs chiefly between the ages of thirty and fifty-five, although it may begin in childhood as the result of congenital syphilis. An overwhelming majority of the cases are in married people, and not infrequently both husband and wife are affected, or one has paresis and the other tabes. Statistics show that it is more common in the lower classes of society, but in America in general medical practice the disease is certainly more common in the well-to-do classes.

**Morbid Anatomy.**—The dura is often thickened, and its inner surface may show the various forms of hypertrophic pachymeningitis. The pia is cloudy, thickened, and adherent to the cortex. The cerebro-spinal fluid is increased in the meningeal spaces, especially in the meshes of the pia, and at times to such an extent as to resemble cysts. The brain is small, and weighs less than normal. The convolutions are atrophied, especially in the anterior and middle lobes. In acute cases the brain may be swollen, hyperæmic, and œdematous. The brain cortex is usually red, and, except in advanced cases, it may not be atrophied, the atrophy of the hemispheres being at the expense of the white matter. The lateral ventricles are dilated to compensate for the atrophy of the brain, and the ependyma may be granular. The fourth ventricle is more constantly dilated, with granulations of its floor covering the calamus scriptorius, a condition seldom seen in any other affection.

In many cases changes are present in the spinal cord and peripheral nerves. There are the typical tabetic changes. There may be degeneration of the pyramidal systems of fibres secondary to the cortical changes. Most com-

monly there is a combination of these two processes. Foci of hæmorrhage and softening, dependent upon coarse vascular changes, are not infrequently found, but are not typical of the disease.

There are various views as to the nature of the changes. The vascular theory is that from an inflammatory process starting in the sheaths of the arterioles there is a diffuse parenchymatous degeneration with atrophic changes in the nerve cells and neuroglia. The syphilitic toxin causes degeneration in the nervous tissues with secondary changes in the neuroglia and vascular system. The spirochætes are found in the brain tissue and rarely in the cord.

**Symptom.**—**PRODROMAL STAGE.**—Irritability, inattention to business amounting sometimes to indifference or apathy, and sometimes a *change in character*, marked by acts which may astonish the friends and relatives, are usually the first indications. There may be unaccountable fatigue after moderate physical or mental exertion. Instead of apathy or indifference there may be an extraordinary degree of physical and mental restlessness. The patient is continually planning and scheming, or may launch into extravagances and speculation of the wildest character. A common feature at this period is the display of an unbounded egoism. He boasts of his personal attainments, his property, his position in life, or of his wife and children. Following these features are important indications of moral perversion, manifested in offences against decency or the law, many of which acts have about them a suspicious effrontery. Forgetfulness is common, and may be shown in inattention to business details and in the minor courtesies of life. At this period there may be no motor phenomena. The onset is usually insidious, although in some cases epileptiform or apoplectiform seizures are the first symptoms. Attacks of hemicrania, like ophthalmic migraine, may occur. Among the early motor features are tremor of the tongue and lips in speaking, slowness of speech, and hesitancy with mixing of syllables or letters. Inequality of the pupils, temporary paresis of the eye muscles with diplopia the Argyll-Robertson pupil, optic atrophy, and changes in the deep reflexes may precede the mental symptoms for years.

**SECOND STAGE.**—This is characterized in brief by mental exaltation or excitement and a progress in the motor symptoms. "The intensity of the excitement is often extreme, acute maniacal states are frequent; incessant restlessness, obstinate sleeplessness, noisy, boisterous excitement, and blind, uncalculating violence especially characterize such states" (Lewis). It is at this stage that the delusion of grandeur becomes marked and the patient believes himself to be possessed of countless millions or to have reached the most exalted sphere possible in profession or occupation. This expansive delirium is, however, not characteristic of general paresis. Besides, it does not always occur, but in its stead there may be marked melancholia or hypochondriasis, or, in other instances, alternate attacks of delirium and depression.

The *facies* has a peculiar stolidity, and in speaking there is marked tremulousness of the lips and facial muscles. The tongue is also tremulous, and may be protruded with difficulty. The speech is slow, interrupted, and blurred. Writing becomes difficult on account of unsteadiness of the hand. Letters, syllables, and words may be omitted. The subject matter of the patient's letters gives valuable indications of the mental condition. In many instances the

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pupils are unequal, irregular, sluggish, sometimes large. Important features in this stage are apoplectiform seizures and paralysis. There may be slight syncopal attacks in which the patient turns pale and may fall. Some of these are *petit mal*. In the apoplectiform seizure the patient falls suddenly, becomes unconscious, the limbs are relaxed, the face is flushed, the breathing stertorous, the temperature increased, and death may occur. Epileptiform seizures are more common than apoplectiform. There may be a definite aura. The attack usually begins on one side and may not spread. There may be twitchings either in the facial or brachial muscles. Typical Jacksonian epilepsy may occur. Recurring attacks of *aphasia* are not uncommon, and paralysis, either monoplegic or hemiplegic, may follow the seizures, or may come on with great suddenness and be transient. In this stage the gait becomes impaired, the patient trips readily, has difficulty in going up or down stairs, and the walk may be spastic or occasionally tabetic. This paresis may be progressive. The deep reflexes are usually increased, but may be lost. Bladder or rectal symptoms gradually develop. The patient becomes helpless, bedridden, and completely demented, and unless care is taken may suffer from bedsores. Death occurs from exhaustion or some intercurrent affection. Spinal cord features may come on with or precede the mental troubles. There are cases in which one is in doubt for a time whether the symptoms indicate *tabes* or general paresis, and it is well to bear in mind that every feature of pre-ataxic *tabes* may exist in the early stage of general paresis.

*Cerebro-Spinal Fluid*.—The findings are as follows: (1) *Cell content*. A lymphocytosis is present in 98-100 per cent. and the average content is 30-60 cells. (2) *Globulin*. This is practically always positive. (3) *Wassermann reaction*. This is positive in nearly every case and usually there is a strong reaction with small amounts. The blood reaction is positive in 98-100 per cent. (4) *Colloidal Gold reaction*. This is nearly always positive in 98-100 per cent., with a typical paretic curve.

### *Tabo-paralysis*

Emphasis has been laid on the identity of the processes underlying *tabes* and general paresis, the spinal cord in the first case receiving the full force of the attack, and the brain in the second. It is suggested that stress determines the location of the process; men whose occupations require much bodily exercise are apt to have *tabes*, while those whose activities are largely mental would suffer from paresis. Usually when the cord symptoms are pronounced the symptoms from the brain remain in abeyance, and the reverse is also true. There are exceptions, and cases of well marked *tabes* may later show the typical symptoms of paresis, but even then the ataxia, if it is not of too high a grade, may improve.

*Optic atrophy*, when it occurs in the pre-ataxic stage of *tabes*, usually indicates that the ataxia will never be pronounced, but unfortunately it is frequently followed by the occurrence of mental symptoms. Mott states that about 50 per cent. of his asylum cases of *tabo-paralysis* had preceding optic atrophy. Its occurrence is therefore of grave significance. The mental symptoms may be delayed for many years.

Made up of a combination of features of the two conditions, the *symptom-*

*complex* of tabo-paralysis varies greatly. It may begin as tabes with lightning pains, bladder symptoms, Argyll-Robertson pupil, loss of the deep reflexes, etc., to have the mental symptoms added later; or, on the other hand, cord symptoms may come on after the patient has shown marked mental changes. The symptoms from the first may be so combined that the name tabo-paralysis is at once applicable. Absent knee-jerks, ocular palsies, or pupillary symptoms may precede the breakdown for many years, but none of them have so grave a significance in regard to the mental state as has optic atrophy. Other types of alienation may interrupt the course of tabes, and the mistake must not be made of regarding them all as general paresis.

**Diagnosis.**—The recognition of general paresis in the earliest stage is extremely difficult, as it is often impossible to decide that the slight alteration in conduct is anything more than one of the moods or phases to which most men are at times subject. The following description by Folsom is an admirable presentation of the diagnostic characters of the early stage: "It should arouse suspicion if, for instance, a strong, healthy man, in or near the prime of life, distinctly not of the 'nervous,' neurotic, or neurasthenic type, shows some loss of interest in his affairs or impaired faculty of attending to them; if he becomes varyingly absent-minded, heedless, indifferent, negligent, apathetic, inconsiderate, and, although able to follow his routine duties, his ability to take up new work is, no matter how little, diminished; if he can less well command mental attention and concentration, conception, perception, reflection, judgment; if there is an unwonted lack of initiative, and if exertion causes unwonted mental and physical fatigue; if the emotions are intensified and easily change, or are excited from trifling causes; if the sexual instinct is not reasonably controlled; if the finer feelings are even slightly blunted; if the person regards with a placid apathy his own acts of indifference and irritability and their consequences, and especially if at times he sees himself in his true light and suddenly fails again to do so; if any symptoms of cerebral vaso-motor disturbances are noticed, however vague or variable."

There are cases of *cerebral syphilis* which closely simulate general paresis. The mode of onset is important, particularly since paralytic symptoms are usually early in syphilis. The affection of the speech and tongue is not present. Epileptic seizures are more common and more liable to be cortical or Jacksonian in character. The expansive delirium is rare. While symptoms of general paresis are not common in connection with gummata or definite gummatous meningitis, paresis may follow closely upon the syphilitic infection. Post mortem in such cases there may be nothing more than a general arterio-sclerosis and diffuse meningo-encephalitis, which may present nothing distinctive. Cases occur in which typical syphilitic lesions are combined with the ordinary lesions of general paresis. There are certain forms of lead encephalopathy which resemble general paresis. *Tumor* may sometimes simulate progressive paresis, but in the former the signs of general increase of the intracranial pressure are usually present. The findings in the spinal fluid are important aids.

**Cytodiagnosis.**—The study of the cerebro-spinal fluid is an important diagnostic measure, particularly in tabes and paresis. Spinal lymphocytosis is the rule and is usually associated with a marked globulin reaction—the normal fluid containing at most minute traces, and a negligible number of cells.

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It is the expression of a subacute or chronic inflammatory process, just as polymorphonuclear leukocytosis is characteristic of an acute process. The syphilitic triad—tabes, paresis, and cerebro-spinal lues—is suggested by lymphocytosis in the spinal fluid. Positive reactions, cytological and chemical, are among the earliest somatic signs, and may clear up obscure cases of tabes and paresis, at the time when diagnosis is most difficult.

**Prognosis.**—The disease rarely ends in recovery. As a rule the progress is slowly downward and the case terminates in a few years, although it is occasionally prolonged ten or fifteen years. There may be remissions in which the patient is able to resume his occupation.

**Treatment.**—Specific treatment has been disappointing on the whole and some authorities regard it as contra-indicated. Certainly some patients are harmed by it but in such a serious condition and as cerebral syphilis may closely resemble it, specific treatment should be tried. The weight of opinion is against intra-spinal treatment. Two measures have been used, the results of which are encouraging. One is infecting the patient with malarial parasites by the injection of blood from a malarial patient. The malaria is allowed to progress for a time and the patient is then given quinine. The other is the use of tryparasamid which has been used in trypanosomiasis. In patients with retinal changes this drug should not be used. The method used by Lorenz and his associates is to give 3 gm. of tryparasamid in 10 c. c. of sterile freshly distilled water intravenously once a week for eight doses. Mercury salicylate (gr. i, 0.06 gm.) is given three days before each dose of the other drug. After five to eight weeks a second similar course is given and if necessary a third course. The serological findings are carefully followed. Retinal changes should be watched for and the drug stopped if there is disturbance of vision. Careful nursing and the orderly life of an asylum are necessary in a great majority of the cases. For sleeplessness and the epileptic seizures bromides may be used. Prolonged remissions, which are not uncommon, are often erroneously attributed to the action of remedies.

### III. DISEASES OF THE EFFERENT OR MOTOR TRACT

#### I. PROGRESSIVE (CENTRAL) MUSCULAR ATROPHY

(*Poliomyelitis anterior chronica; Amyotrophic Lateral Sclerosis; Progressive Bulbar Paralysis*)

**Definition.**—A disease characterized by a chronic degeneration of the motor tract, usually of the whole, but at times limited to the lower segment. Associated with it is a progressive atrophy of the muscles, with more or less spastic rigidity.

Three affections, as a rule described apart, belong together in this category: (a) Progressive muscular atrophy of spinal origin; (b) amyotrophic lateral sclerosis; and (c) progressive bulbar paralysis. A slow atrophic change in the motor neurones is the anatomical basis, and the disease involves, in many cases, the cortical, bulbar, and spinal centres. There may be simple muscular

atrophy with little or no spasm, or progressive wasting with marked spasm and great increase in the reflexes. There may be symptoms of involvement of the motor nuclei in the medulla—a glosso-labio-laryngeal paralysis; while in others, again, with atrophy (especially of the arms) a spastic condition of the legs and bulbar phenomena, tremors develop and signs of cortical lesion. These various stages may be traced in the same case.

For convenience, bulbar paralysis is considered separately, and *progressive muscular atrophy* and *amyotrophic lateral sclerosis* are taken together.

**History.**—The disease is known as the Aran-Duchenne type of progressive muscular atrophy and as Cruveilhier's palsy, after the French physicians who described it. Luys and Lockhart Clarke first demonstrated that the cells of the ventral horns of the spinal cord were diseased. Charcot separated two types—one with simple wasting of the muscles, due, he believed, to degeneration confined to the ventral horns (and to this he restricted the name progressive muscular atrophy—type, Aran-Duchenne); the other, in which there was spastic paralysis of the muscles followed by atrophy. As the anatomical basis for this he assumed a primary degeneration of the pyramidal tracts and a secondary atrophy of the ventral horns. To this he gave the name of amyotrophic lateral sclerosis. There is but little evidence to show that any such sharp distinction can be made between these two diseases.

**Etiology.**—The cause is doubtful. Syphilis is associated with some of the cases and this form of atrophy has occurred in tabes dorsalis. It is more frequent in males than in females and affects adults, usually after the thirtieth year, though occasionally younger persons are attacked. Cases of progressive muscular atrophy in early life belong as a rule to the dystrophies. Certain cases follow injury. The Werdnig-Hoffman type is a familial affection and does not belong here. The spastic form may develop late in life—after seventy—as a senile change.

**Morbid Anatomy.**—The essential anatomical change is a slow degeneration of the motor path, involving particularly the lower motor neurones. The upper neurones are also involved, either first, simultaneously, or at a later period. Associated with the degeneration in the cells of the ventral horns there is a degenerative atrophy of the muscles. The following are the important anatomical changes: (a) The gray matter of the cord shows the most marked alteration. The large ganglion cells of the ventral horns are atrophied, or, in places, have entirely disappeared, the neuroglia is increased, and the medullated fibres are much decreased. The fibres of the ventral nerve-roots passing through the white matter are wasted. (b) The ventral roots outside of the cord are also atrophied. (c) The muscles affected show degenerative atrophy, and the inter-muscular branches of the motor nerves are degenerated. (d) The degeneration of the gray matter is rarely confined to the cord, but extends to the medulla, where the nuclei of the motor cerebral nerves are extensively wasted. (e) In a majority of the cases there is sclerosis in the ventro-lateral white tracts, the lateral pyramidal tracts particularly are diseased, but the degeneration is not confined to them, but extends into the ventro-lateral ground bundles. The direct cerebellar and the ventro-lateral ascending tracts are spared. The degeneration in the pyramidal tracts extends to the brain to the motor cortex, the cells of which are degenerated. In the medulla the medial longitudinal fasciculus has been found diseased. (f) In

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those cases in which no sclerosis has been found in the pyramidal tracts there has been a sclerosis of the ventro-lateral ground bundle (short tracts).

**Symptoms.**—Irregular pains, sensory disturbances or feelings of stiffness may precede the onset of the wasting which is usually gradual. The hands are usually first affected, and there is difficulty in performing delicate manipulations. The muscles of the ball of the thumb waste early, then the interossei and lumbricales, leaving marked depressions between the metacarpal bones. Ultimately the contraction of the flexor and extensor muscles and the extreme atrophy of the thumb muscles, the interossei, and lumbricales produce the claw-hand—*main en griffe* of Duchenne. The flexors of the forearm are usually involved before the extensors. In the shoulder-girdle the deltoid is first affected; it may waste before the other muscles of the upper extremity. The trunk muscles are gradually attacked; the upper part of the trapezius long remains unaffected. Owing to the feebleness of the muscles which support it, the head tends to fall forward. The platysma myoides is unaffected and often hypertrophies. The arms and the trunk muscles may be much atrophied before the legs are attacked. The face muscles are usually attacked late.

Ultimately the intercostal and abdominal muscles may be involved, the wasting proceeds to an extreme grade, and the patient may be actually "skin and bone," and, as "living skeletons," the cases are not uncommon in "museums" and "side-shows." Deformities and contractures result, and lordosis is almost always present. A curious twitching of the muscles (fibrillation) is common and may occur in muscles which are not attacked. It is an important sign but is not a characteristic feature. The irritability of the muscles is increased. Sensation is unimpaired, but the patient may complain of numbness and coldness of the affected limbs. The galvanic and faradic irritability of the muscles progressively diminishes and may become extinct, the galvanic persisting for the longer time. In cases of rapid wasting and paralysis the reaction of degeneration may be obtained. The excitability of the nerve trunks may persist after the muscles have ceased to respond. The loss of power is usually proportionate to the wasting.

**Amyotrophic Spastic Form.**—The foregoing description applies to the group of cases in which the atrophy and paralysis are flaccid—*atonic*, as Gowers called it. In other cases those which Charcot described as amyotrophic lateral sclerosis, spastic paralysis precedes the wasting. The reflexes are greatly increased. It is one of the rare conditions in which a jaw clonus may be obtained. The most typical condition of spastic paraplegia may be produced. On starting to walk, the patient seems glued to the ground and makes ineffectual attempts to lift the toes; then four or five short, quick steps are taken on the toes with the body thrown forward; and finally he starts off, sometimes with great rapidity. Some of the patients can walk up and down stairs better than on the level. The wasting is never so extreme as in the atonic form, and the loss of power may be out of proportion to it. The sphincters are unaffected. Sexual power may be lost early. A flaccid atrophic paralysis with increased reflexes is the common finding. The differences depend upon the relative extent of involvement of the upper and lower motor segments and the time of the involvement of each. The condition may be unilateral.

As the degeneration extends upward an important change takes place from the occurrence of bulbar symptoms, which may, however, precede the spinal manifestations. The lips, tongue, face, pharynx, and larynx may be involved. The lips may be affected and articulation impaired for years before serious symptoms occur. In the final stage there may be tremor, the memory fails, and a condition of dementia supervenes.

**Diagnosis.**—Progressive (central) muscular atrophy begins, as a rule, in adult life, without hereditary or family influences (the early infantile form being an exception), and usually affects first the muscles of the thumb, and gradually involves the interossei and lumbricales. Fibrillary contractions are common, electrical changes occur, and the deep reflexes are usually increased. These are usually sufficient to distinguish it from the other forms of muscular wasting. It is well to remember that the earliest and most marked indication of *cervical rib* may be atrophy of the small muscles of the hand. In *syringomyelia* the sensory disturbances, as a rule, make the diagnosis clear, but when these are absent or but little developed it may be very difficult to distinguish the disease. Neoplasm of the cord or spine or pachymeningitis may give difficulty but a careful study, especially of the spinal fluid, observation for a short time and the absence of fibrillation should prevent error.

**Treatment.**—The disease is incurable. The downward progress is slow but certain, though in a few cases a temporary arrest may take place. Arsenic and hypodermic injections of strychnine may be tried. If syphilis is present, specific treatment does not result in much benefit. Systematic massage is useful in the spastic cases.

#### *Bulbar Paralysis (Glosso-labio-laryngeal Paralysis)*

When the disease affects the motor nuclei of the medulla first or early, it is called bulbar paralysis, but it has practically no independent existence, as the spinal cord is sooner or later involved.

**Symptoms.**—The disease begins with slight defect in the speech, and difficulty in pronouncing the dentals and linguals. The paralysis starts in the tongue, and the superior lingual muscle gradually becomes atrophied, and finally the mucous membrane is thrown into transverse folds. In the process of wasting the fibrillary tremors are seen. Owing to the loss of power in the tongue, the food is with difficulty pushed back into the pharynx. The saliva also may be increased, and is apt to accumulate in the mouth. When the lips become involved the patient can neither whistle nor pronounce the labial consonants. The mouth looks large, the lips are prominent, and there is constant drooling. The food is masticated with difficulty. Swallowing becomes difficult, owing partly to the regurgitation into the nostrils, partly to the involvement of the pharyngeal muscles. The muscles of the vocal cords waste and the voice becomes feeble, but the laryngeal paralysis is rarely so extreme as that of the lips and tongue.

The **course** is slow but progressive. Death may result from an aspiration pneumonia, sometimes from choking, more rarely from involvement of the respiratory centres. The mind usually remains clear. The patient may become emotional. In a majority of the cases the disease is only part of a progressive atrophy, either simple or associated with a spastic condition. In

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the later stage of amyotrophic lateral sclerosis the bulbar lesions may paralyze the lips long before the pharynx or larynx becomes affected.

The **diagnosis** is readily made, either in the acute or chronic form. The involvement of the lips and tongue is usually well marked, while that of the palate may be long deferred. In *pseudo-bulbar paralysis* bilateral disease of the motor cortex in the lower part of the ascending frontal convolution, or about the knee of the internal capsule may interfere with the supranuclear paths, causing paralysis of the lips and tongue and pharynx which closely simulates a lesion of the medulla. Sometimes the symptoms appear on one side, but they may develop suddenly on both sides. Bilateral lesions have usually been found, but the disease may be unilateral. There is arteriosclerosis and the bulbar features are usually sequels of hemiplegic attacks.

*Acute bulbar paralysis* may be due to (a) hæmorrhagic or embolic softening in the pons and medulla; (b) acute inflammatory softening, analogous to polio-myelitis, occurring occasionally as a post-febrile affection. It has occasionally followed diphtheria, and occurred after severe electric shocks of high voltage. It usually comes on very suddenly and the symptoms may correspond closely to those of an advanced case of chronic bulbar paralysis. The sudden onset and the associated symptoms make the diagnosis easy. In these acute cases there may be loss of power in one arm, or hemiplegia, sometimes alternate hemiplegia, with paralysis on one side of the face and loss of power on the other side of the body. (c) In polio-myelitis and epidemic encephalitis there are cases with acute bulbar symptoms.

## II. SPASTIC PARALYSIS OF ADULTS

### (Primary Lateral Sclerosis)

**Definition.**—A gradual loss of power with spasm of the muscles of the body, the lower extremities being first and most affected, unaccompanied by muscular atrophy, sensory disturbance, or other symptoms. A systemic degeneration of the pyramidal tracts is assumed.

**Symptoms.**—The general symptoms of spastic paraplegia in adults are very distinctive. The patient complains of feeling tired, of stiffness in the legs, and perhaps of pains of a dull aching character in the back or in the calves. There may be no definite loss of power, even when the spastic condition is well established. In other instances there is definite weakness. The stiffness is felt most in the morning. In a well developed case the gait is most characteristic. The legs are moved stiffly and with hesitation, the toes drag and catch against the ground, and, in extreme cases, when the ball of the foot rests upon the ground a distinct clonus develops. The legs are kept close together, the knees touch, and in certain cases the adductor spasm may cause cross-legged progression. On examination, the legs may at first appear tolerably supple, perhaps flexed and extended readily. In other cases the rigidity is marked, particularly when the limbs are extended. The spasm of the adductors of the thigh may be so extreme that the legs are separated with the greatest difficulty. In cases of this extreme rigidity the patient usually loses the power of walking. The nutrition is well maintained, the muscles may be hypertrophied. The reflexes are greatly increased. The slightest

touch upon the patellar tendon produces an active knee-jerk. The rectus clonus and the ankle clonus are easily obtained. In some instances the slightest touch may throw the legs into violent clonic spasm, the condition to which Brown-Séquard gave the name of spinal epilepsy. The superficial reflexes are also increased. The arms may be unaffected for years, but occasionally they become weak and stiff at the same time as the legs.

The course of the disease is progressively downward. Years may elapse before the patient is bedridden. Involvement of the sphincters, as a rule, is late; occasionally it is early. The sensory symptoms rarely progress, and the patients may retain their general nutrition and enjoy excellent health. Ocular symptoms are rare.

**Diagnosis.**—The diagnosis, so far as the clinical picture is concerned, is readily made, but it is often very difficult to determine accurately the nature of the underlying pathological condition. A history of syphilis is present in many of the cases. Cases which have run a fairly typical clinical course upon coming to autopsy have been found to have been due to very different conditions—transverse myelitis, multiple sclerosis, cerebral tumor, etc. General paresis may begin with symptoms of spastic paraplegia, and Westphal believed that it was only in relation to this disease that a primary sclerosis of the pyramidal tracts ever occurred. In any case the diagnosis of primary systemic degeneration of the pyramidal tract is, to say the least, doubtful.

**Treatment.**—Not much can be done to check the progress. Division of the posterior nerve roots is permissible when the motor weakness is due chiefly to spasticity. A number of patients have been operated upon successfully. The same has been done in the spasticity with bilateral athetosis.

### III. SECONDARY SPASTIC PARALYSIS

Following any lesion of the pyramidal tract there may be a spastic paralysis; thus, in a transverse lesion of the cord, whether the result of slow compression (as in caries), chronic myelitis, the pressure of tumor, chronic meningo-myelitis, or multiple sclerosis, degeneration takes place in the pyramidal tracts, below the point of disease. The legs soon become stiff and rigid, and the reflexes increase. Bastian has shown that in compression paraplegia if the transverse lesion is complete, the limbs may be flaccid, without increase in the reflexes—*paraplégie flasque* of the French. The condition of the patient in these secondary forms varies very much. In chronic myelitis or in multiple sclerosis he may be able to walk about, but with a characteristic spastic gait. In the compression myelitis, in fracture, or in caries, there may be complete loss of power with rigidity.

It may be difficult or even impossible to distinguish these cases from those of primary spastic paralysis. Reliance is to be placed upon the associated symptoms; when these are absent no definite diagnosis as to the cause of the spastic paralysis can be given.

**Syphilitic Spinal Paralysis.**—Erb described a symptom group under the term syphilitic spinal paralysis. The points upon which he laid stress are a very gradual onset with a development finally of the features of a spastic paresis; the tendon reflexes are increased, but the muscular rigidity is slight in comparison with the exaggerated deep reflexes. There is rarely much pain,

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and the sensory disturbances are trivial, but there may be paræsthesia and the girdle sensation. The bladder and rectum are usually involved, and there is sexual failure or impotence. And, lastly, improvement is not infrequent. A majority of instances of spastic paralysis of adults not the result of slow compression of the cord are associated with syphilis and belong to this group.

### C. HEREDITARY AND FAMILIAL DISEASES

#### I. THE MUSCULAR DYSTROPHIES

(*Dystrophia muscularis progressiva*, Erb. *Primary Myopathy*)

**Definition.**—Muscular wasting, with or without an initial hypertrophy, beginning in various groups of muscles, usually progressive in character, and dependent on primary changes in the muscles themselves or the neuro-muscular endings.

**Etiology.**—No factor other than heredity is known, which may show itself by true heredity—the disease occurring in two or more generations—or several members of the same generation may be affected. Members of the same family may be attacked through several generations: as many as 20 or 30 cases have been described in five generations. Males, as a rule, are more frequently affected than females. In families, persons of the same sex are usually attacked, but unaffected females may transmit the disease. In Erb's cases 44 per cent. showed no heredity. The disease usually sets in before puberty, but the onset may be as late as the twenty-fifth year, or even later.

**Pathology.**—At first the muscle fibres hypertrophy, and become round; the nuclei increase, and the fibres may become fissured. At the same time there is a slight increase in the connective tissue. Sooner or later the fibres begin to atrophy, and the nuclei become greatly increased. Vacuoles and fissures appear, and the fibres become completely atrophic, the connective tissue increasing with deposition of fat to such an extent as to cause hypertrophic lipomatosis—pseudo-hypertrophy. The different stages of these changes may be found in a single muscle at the same time.

The nervous system has very generally been found to be without demonstrable lesions, but in certain cases changes in the cells of the ventral horns have been described. Changes in the *pineal gland*, producing shadows, have been demonstrated by the X-rays, from which very naturally it is suggested that the disease is due to a disturbance in the internal secretions.

**Symptoms.**—Clumsiness in the movements of the child is the first symptom noticed and on examination certain muscles or groups of muscles seem to be enlarged, particularly those of the calves. The extensors of the leg, the glutei, the lumbar muscles, the deltoid, triceps and infraspinatus, are the next most frequently involved, and may stand out with great prominence. The muscles of the neck, face, and forearm rarely suffer. Sometimes only a portion of a muscle is involved. With this hypertrophy of some muscles there is wasting of others, particularly the lower portion of the pectorals and the latissimus dorsi. The attitude when standing is very characteristic. The

are far apart, the shoulders thrown back, the spine is greatly curved, and the abdomen protrudes. The gait is waddling and awkward. In getting up from the floor the position assumed, so well known by Gowers' figures, is pathognomonic. The patient first turns over in the all-fours position and raises the trunk with his arms; the hands are then moved along the ground until the knees are reached; then with one hand upon a knee he lifts himself up, grasps the other knee, and gradually pushes himself in the erect posture, as it has been expressed, by climbing up his legs. The striking contrast between the feebleness of the child and the powerful looking pseudo-hypertrophic muscles is very characteristic. The enlarged muscles may, however, be relatively very strong.

The course is slow, but progressive and usually not over ten years. Wasting proceeds and finally all traces of the enlarged condition of the muscles disappear. At this late period distortions and contractions are common. The muscles of the shoulder-girdle are nearly always affected early, causing a symptom upon which Erb lays great stress. With the hands under the arms, when one endeavors to lift the patient, the shoulders are raised to the level of the ears, and one gets the impression as though the child were slipping through. These "loose shoulders" are very characteristic. The abnormal mobility of the shoulder blades gives them a winged appearance, and makes the arms seem much longer than usual when they are stretched out.

There are no sensory symptoms. The atrophic muscles do not show the reaction of degeneration except in extremely rare instances.

**Clinical Forms.**—A number of forms have been described, depending upon the age at onset, the muscles first affected, the occurrence of hypertrophy, heredity, etc., but there is no sharp division between the forms. The following are the more important:

I. *The pseudo-hypertrophic type* of Duchenne, most common in childhood and in family groups. The hypertrophy of the muscles is the striking feature, whether a true hypertrophy or a lipomatosis. There is also a juvenile type with atrophy, affecting chiefly the shoulder girdles and upper arms. Isolated cases occur in adults.

II. *The facio-scapulo-humeral type* of Landouzy-Déjerine. The face is first involved, causing the myopathic facies, the lips prominent, the upper one projecting, the eyes cannot be closed, nor the forehead wrinkled, the smile is transverse, from inaction of the levators of the lip. Later the shoulder-girdle muscles are involved, the scapulae are winged, the upper arms wasted, and lastly, the thigh muscles. With all this there may be no hypertrophy, though often, if carefully sought, there will be found areas of enlargement—the so-called muscle balls. This form may begin in adults.

III. *The thigh-muscle type* of Leyden, Moebius and Zimmerlin, in which the disease starts in the extensors of the thighs which are deeply involved before other groups of upper arms and trunk are attacked.

In all forms, when the muscles of the trunk become involved, there is flattening of the chest and the peculiar "wasp-waist" described by Marie.

**Diagnosis.**—The muscular dystrophies can usually be distinguished readily from the other forms of muscular atrophy.

(a) In the cerebral atrophy loss of power usually precedes the atrophy.

(b) Progressive (central) muscular atrophy begins in the small muscles

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of the hand, the reaction of degeneration is present and fibrillary twitchings occur in both the atrophied and non-atrophied muscles. The central atrophies come late in life, the dystrophies, as a rule, early. In the progressive muscular dystrophies heredity plays an important rôle. In the rare cases of early infantile spinal muscular atrophy occurring in families the symptoms are so characteristic of a central disease that the diagnosis presents no difficulty.

(c) In the neuritic muscular atrophies, due to lead or trauma, seen for the first time when the wasting is marked there is often difficulty, but the absence of family history and the distribution are important, while the paralysis is out of proportion to the atrophy. Sensory symptoms may be present.

(d) Progressive neural muscular atrophy. Here heredity is also a factor, and the disease usually begins in early life, but the distribution of atrophy and paralysis, which is at first confined to the periphery of the extremities, helps to distinguish it from the dystrophies.

**Prognosis.**—The outlook in the primary muscular dystrophies is bad. The wasting progresses uniformly, uninfluenced by treatment.

**Treatment.**—By electricity and massage the progress may be occasionally retarded. The general health should be carefully looked after, moderate exercise allowed with friction of the muscles with oil, and when the patient becomes bedfast, as is inevitable sooner or later, care should be taken to prevent contractures in awkward positions.

### II. FAMILIAL SPINAL MUSCULAR ATROPHY

(*Werdnig-Hoffman*)

A rare disease which may be hereditary as well as occurring in a family without disease in the ascendants. Anatomically there is marked degeneration of the anterior horns in the spinal cord, of the anterior roots, and less marked changes in the peripheral nerves, with widespread atrophy of the muscular fibres. While in many cases the disease resembles muscular dystrophy, anatomically it appears to be a progressive central muscular atrophy. The onset is in the first year, sometimes prenatal, often within two months of birth. There is weakness of the trunk muscles and later in the limbs. The paralysis is followed by atrophy of the muscles which may show fibrillary tremors. The trunk muscles finally become completely paralyzed and death may result from paralysis of the intercostal muscles and diaphragm or from bulbar paralysis. The course is progressive and the later the onset the longer the course. In the prenatal cases or those beginning soon after birth the course is rapid. Death usually occurs within a year. The *diagnosis* from amyotonia congenita may be difficult but in this the paralysis is not complete and tends to improve. There is no treatment of any value.

### III. PROGRESSIVE NEURAL MUSCULAR ATROPHY

(*Peroneal Type and Hypertrophic Type—Charcot-Marie-Tooth*)

The peroneal type, described first by Charcot, Marie, and Tooth, is a hereditary and familial disease beginning in childhood, affecting first the

muscles of the peroneal group, leading to club-foot. The disease seems to occupy a position between central muscular atrophy and the muscular dystrophies, resembling the latter in the early onset and familial character, and the former in the occurrence of fibrillary twitchings, the presence of electrical changes and the implication of the small muscles of the hand. Anatomically sclerosis of the posterior and postero-lateral columns, atrophy of the cells of the anterior horns and alterations of the peripheral nerves are found.

The essential feature is implication of the distal with normal proximal portions of the limbs, which gives a very characteristic picture. There is great decrease of the electrical excitability. Ocular symptoms are rare; occasionally there is atrophy of the optic nerves. The disease should be suspected in cases of acquired double club-foot.

#### IV. PROGRESSIVE INTERSTITIAL HYPERTROPHIC NEURITIS

This is a familial disease beginning, as a rule, in infancy with a combination of the symptoms of tabes and muscular atrophy. Anatomically there is sclerosis of the posterior columns of the cord with interstitial hypertrophic neuritis. It was first described by Déjerine and Sottas, and, though rare, a good many families have been reported, one by Marie in which seven children were affected. The spinal cord lesions resemble those of tabes, and result from degeneration of the posterior nerve roots. The hypertrophy of the nerves is of a unique type. The affected muscles waste completely and there is replacement by fibrous tissue.

The *symptoms* begin in early life and are: (a) Incoördination very like that of tabes dorsalis, only as the disease progresses the gait is steppage; (b) sensory disturbances, sometimes pains which are fulgurant in character; (c) muscular atrophy, limbs and face, in the former chiefly distal and in the legs extending up to the lower third of the thigh. The feet are usually in the varus position; kypho-scoliosis is also present. (d) Ocular symptoms are marked—myosis, Argyll-Robertson sign. (e) The peripheral nerves are hypertrophied, sometimes double the normal size, smooth and not painful, those of the lower limbs being chiefly involved. The optic and olfactory nerves escape. The peculiar distribution usually gives the diagnosis. The disease should be suspected in acquired double club-foot. In *treatment* exercises should be used suitable for the strength; fatigue should be avoided. Massage and electricity are useful. Heavy apparatus should be avoided and tenotomy not done. Light splints should be worn at night to lessen the deformity.

#### V. HEREDITARY ATAXIA

(Friedreich's Ataxia)

**Definition.**—A familial disease occurring in childhood characterized by locomotor and static ataxia, speech disturbances and nystagmus, and anatomically by degeneration of the postero-lateral and spino-cerebellar tracts. In 1863 Friedreich first reported six cases.

**Etiology.**—It is a family disease; the 143 cases analyzed by Griffiths occurred in 71 unrelated families. Males are most frequently attacked, 86 to

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57 in Griffith's series. Direct inheritance occurs and was noted in 33 cases. The onset is usually before puberty, but may be as late as the 25th year. The cause is unknown. Various influences in the parents, such as consanguinity, alcoholism, and syphilis have been reported. The disease belongs to Gower's abiotrophies, an inherited weakness, lack of vitality in certain sections of the nervous system, leading to early degeneration.

**Morbid Anatomy.**—Both cord and cerebellum have been reported smaller than usual. The posterior meninges may be thickened. The important change is a complete sclerotic degeneration of the postero-lateral tracts forming the most typical example of combined degeneration. Gowers' tract and the direct cerebellar are always involved. Déjerine and Letulle suggest that the disease differs from ordinary spinal sclerosis and is a gliosis due to developmental errors.

**Symptoms.**—The inco-ordination begins in the legs, and the gait is swaying, irregular, and more like that of a drunken man without the characteristic stamping gait of the true tabes. Romberg's sign may or may not be present. The ataxia of the arms occurs early and is very marked; the movements are almost choreiform, irregular and somewhat swaying. In making any voluntary movement the action is overdone, the prehension is clawlike, and the fingers may be spread or overextended just before grasping an object. The hand frequently moves about an object for a moment, and then suddenly pounces upon it. There are irregular, swaying movements of the head and shoulders. There is present in many cases what is known as *static ataxia*, that is to say, ataxia of quiet action. It occurs when the body is held erect or when a limb is extended—irregular, oscillating movements of the head and body or of the extended limb.

The muscle tone depends on the relative degeneration in the posterior roots and pyramidal tracts; the former tends to abolish and the latter to increase it. The same governs the tendon reflexes; they are usually absent, sometimes early, but if the pyramidal tracts are specially affected the reflexes persist. The plantar reflex shows an extensor response. The skin and eye reflexes are normal. Sensory symptoms are not usually present. The cerebrospinal fluid is normal.

*Nystagmus* is a characteristic feature. Atrophy of the optic nerve rarely occurs. Disturbance of *speech* is common; usually slow and scanning; it may be explosive. The expression is often dull; the mental power is, as a rule, maintained, but late in the disease becomes impaired. A striking feature is early deformity of the feet, so that the patient walks on the outer edge of the feet. The big toe is flexed dorsally on the first phalanx. *Scoliosis* is common.

The course is usually progressive but slow. As the disease advances, paralysis comes on and may ultimately be complete. Some of the patients never walk.

**Diagnosis.**—This is not difficult when several members of a family are affected. The onset in childhood, the curious form of inco-ordination, the loss of knee-jerks, the early deformity of the feet, the position of the great toe, *scoliosis*, *nystagmus*, and speech disturbance make up an unmistakable picture. With hereditary chorea it has certain similarities, but usually this disease does not set in until after the 30th year.

**Treatment.**—The disease is incurable but every effort should be made to keep up the general health. The patients should not be put to bed unless this is absolutely necessary as inactivity often does harm. Comfortable boots for the deformed feet should be worn. Educational training of the muscular movements as in tabes is useful.

## VI. HEREDITARY CEREBELLAR ATAXIA (*Marie*)

This closely resembles Friedreich's ataxia in so far as the ataxia, nystagmus and speech disturbances are concerned. It differs in (1) a later onset (after twenty) in many cases; (2) the ataxia is more purely cerebellar; (3) there is spasticity of the legs with increased knee-jerks; (4) there is no talipes or scoliosis; and (5) optic atrophy and ocular palsies are common with the occasional presence of the Argyll-Robertson pupil. Marie regarded the lesion as atrophy of the cerebellum but the lesions may be more widely spread and resemble the Friedreich form. In the form termed spino-cerebellar or Sanger Brown's ataxia there is degeneration of the spino-cerebellar tracts and less marked change in the dorsal columns. The pyramidal tracts are usually not affected. The ataxia, speech disturbance and movements are like those in the Friedreich form but the onset is later in life, nystagmus is rare, ocular paralyses occur, optic atrophy is common, and scoliosis does not occur.

There are other forms of cerebellar ataxia resembling those discussed. (1) Primary progressive cerebellar ataxia shows degeneration of the cortex cells of the cerebellum and of the connecting fibres with the central nuclei. The cerebellum is reduced in size. The onset is in middle life. Ataxia, nystagmus, speech disturbance and irregular movements occur. The reflexes are normal. (2) In the form described by Déjerine and Thomas the lesions are more widespread, involving the olivary bodies, the pons and cerebellar peduncles. Atrophy of the cerebellum occurs. The onset is usually in middle age. The features are much like the preceding form.

## VII. HEREDITARY SPASTIC PARAPLEGIA

**Definition.**—A familial, abiotrophic, disease, involving chiefly the pyramidal tracts. It is sometimes hereditary.

**Etiology.**—It begins in children, usually after the seventh year; the onset may be delayed until the twentieth: three or four members of a family may be attacked; boys more often than girls in the proportion of 88 to 51 (Deléarde and Minet). In some families in which the disease has been hereditary, the females have escaped. Mild cases in a family may exist with increase of the reflexes as the only symptom.

**Pathology.**—The spinal degeneration is chiefly in the pyramidal tracts of the lumbar and lower thoracic regions. In the late stages the lesions may be those of a combined sclerosis with involvement of the direct cerebellar tracts. Newmark's studies show imperfect development of the cord (agenesia) as an important factor.

**Symptoms.**—Early exaggeration of the knee-jerks may precede any paralysis or weakness; gradually there are spasticity and Babinski's sign, with contractures and paralysis. The abdominal reflexes disappear. It is important

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to rule out the cases with mental features and Little's disease. The paralysis may extend to the upper limbs, and eyes and speech are involved. In others again there is atrophy of the muscles, and the picture is not unlike amyotrophic lateral sclerosis, or a disseminated sclerosis. Very different pictures may be presented by affected children in the same family.

### VIII. CHRONIC HEREDITARY CHOREA

#### *(Huntington's Chorea)*

**Definition.**—A hereditary disease characterized by irregular movements, disturbance of speech and progressive mental deterioration.

**History.**—In 1863 Lyon described it as chronic hereditary chorea. In 1872 George Huntington, whose father, grandfather, and great-grandfather had treated cases, gave in three brief paragraphs its salient features—heredity, the late onset, and the mental changes. The disease is more common in the United States than in Europe. Davenport has studied the four great family complexes of eastern Long Island, southwestern Connecticut, south-central Connecticut, and eastern Massachusetts “which show nearly 1000 cases, of Huntington's chorea, and yielding the remarkable results that practically all can be traced back to some half-dozen individuals, including three (probable) brothers who migrated to America in the XVIIth century.”

**Inheritance.**—It never skips a generation. The age of onset does not appear to vary, averaging from thirty-five to thirty-eight. The mental type is usually hyperkinetic. Among 3000 persons related to the 962 cases studied by Davenport, there were many other nervous disorders—epilepsy in 39, infantile convulsions in 19, and feeble-mindedness in 73.

**Pathology.**—There is marked destruction of the smaller ganglion cells of the globus pallidus system which have a co-ordinating and inhibitory control over the larger motor cells. When this is lost chorea results (Hunt). The large cell system of the globus pallidus stands in relation to the paralysis agitans syndrome and the small cell system to the chorea syndrome. The other findings are varied. Meningeal thickening and atrophy of the cortex, with a loss of cells, have been present in some cases. Arterio-sclerotic changes are common in older subjects.

**Symptoms.**—Difficulty in performing delicate actions with the hands, as in writing, or in buttoning a shirt collar, may be the earliest indication, or there are slight involuntary movements of the head and face. When well established, the movements are slower than in Sydenham's chorea, irregular and incoördinate. The face muscles are early involved, causing involuntary grimaces. The gait is irregular and swaying, not unlike that of a drunken man. The speech is slow and the syllables blurred. The reflexes, not altered at first, are later increased. Certain biotypes have been observed by Davenport. Thus the tremors may be absent and the mental condition present, or the muscular movements may be present without mental defects. The chorea may not progress and the onset may be early in life. He found family differences in all these points.

The mental changes may come early, outbreaks of temper and excitement are common, alternating with periods of depression. Usually a progressive

failure of the mental powers leads to complete dementia. Dreading a terrible fate, it is not surprising to hear of suicide in certain members of the families.

Little or nothing can be done to arrest the progress of the disease.

**Prevention.**—Davenport's study shows how much more serious the disease is than we had hitherto thought. It is transmitted through males and females, and Davenport states that there is no evidence of any abstention from or selection against marrying in the members of the large group of hereditary choreas studied by him. There is no efficient treatment.

## IX. PROGRESSIVE LENTICULAR DEGENERATION

(*Wilson's Disease: Hepato-lenticular Degeneration*)

**Definition.**—A familial, not hereditary, extra-pyramidal disease usually coming on early in life, characterized by tremor and spasticity with bilateral changes in the lenticular nuclei and cirrhosis of the liver.

Described by Wilson in 1912, it is apparently the same condition which Gowers designated tetanoid chorea and resembles the pseudo-sclerosis of Westphal and Strümpell. The onset is between the ages of 10 and 25 years. As to pathogenesis Wilson suggests the selective action of some toxin possibly due to the hepatic cirrhosis. The lenticular nuclei show degeneration with cavitation and atrophy. The process may extend more widely to the internal capsule, motor cortex, basal ganglia and pyramidal tracts. The cirrhosis of the liver is marked and of a mixed type.

The main features are disturbance of motor functions, rigidity, difficulty in deglutition and speech, emotional disturbances and a terminal dementia. There are no true paralyses, sensory symptoms or changes in the reflexes. The involuntary movements may be of the nature of tremor, choreiform or athetoid; they involve the extremities and are often increased on effort. There are muscular rigidity, weakness, spasticity and painful muscular contractions. When the patient grasps an object he may have difficulty in relaxing his hold. As the disease progresses contractures and progressive emaciation are marked. The patient becomes helpless and is unable to use the arms and legs or turn in bed. The hepatic cirrhosis does not seem to have caused any symptoms or signs in the reported cases. A curious annular brownish-green pigmentation of the cornea has been noted in a few cases. The disease is progressive with a course in acute cases of a few months and in chronic forms of two to seven years. There is no specific treatment.

## X. PERIODIC PARALYSIS

**Definition.**—A recurring paralysis, lasting from a few hours to a few days, affecting members of the same family, with abolition of the faradic excitability of both muscles and nerves. Death may occur in an attack.

**History.**—After a few scattered references, the disease was accurately described in 1885 by Westphal and Oppenheim. Family groups then began to be recognized, and now a large number of cases have been studied.

**Etiology.**—The majority have occurred in groups but sporadic cases occur. Holtzapfel reported seventeen cases in four generations. Many members

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of this family suffered from migraine. Transmission is either through the male or female; the disease may skip a generation.

**Pathology.**—Nothing definite is known. Winternitz could find no organic lesions in two fatal cases in the family reported by Holtzapfle. Naturally auto-intoxication has been suggested, and extensive researches into metabolism have been made. Diminution of creatinin excretion has been determined. In some respects the disease is similar to Myasthenia gravis, in which there are attacks of transient paralysis. Westphal regarded the disease as a vasomotor neurosis associated with migraine, which was such a striking feature in Holtzapfle's cases. Temporary collapse of the vessels is met with in this condition, and Holtzapfle suggests that this may occur in the anterior horns.

**Symptoms.**—The clinical picture is similar in all recorded cases. The paralysis involves, as a rule, the arms and legs, but may be general below the neck. It comes on in healthy persons without apparent cause, and often during sleep. At first there may be weakness of the limbs, a feeling of weariness and sleepiness, but rarely with sensory symptoms. The paralysis, beginning in the legs, to which it may be confined, is usually complete within the first twenty-four hours. The neck muscles are sometimes involved, and occasionally those of the tongue and pharynx. The cerebral nerves and the special senses are, as a rule, unaffected. The temperature is normal or subnormal, and the pulse slow. The deep reflexes are diminished, sometimes abolished, and the skin reflexes may be enfeebled. The faradic excitability of both muscles and nerves is reduced or abolished. Improvement begins within a few hours or a day or two, the paralysis disappearing completely and the patient becoming perfectly well. The attacks usually recur at intervals of one to two weeks, but they may return daily. They generally cease after the fiftieth year. There may be signs of acute dilatation of the heart during the attack.

**Treatment.**—A low protein diet is advisable and free elimination should be ensured. Potassium bromide (gr. 30, 2 gm.) with caffeine citrate (gr. ii, 0.13 gm.) has been of value early in the attack and potassium citrate in full doses may shorten or abort an attack.

### XI. AMAUROTIC FAMILY IDIOCY

(*Tay-Sachs' Disease*)

**Definition.**—A family disease of infancy characterized by mental impairment progressing to idiocy, progressive muscular weakness, macular changes in the retina and a fatal termination.

**History.**—In 1881 Waren Tay reported a group of cases characterized by muscular weakness, macular lesions, and death before the age of two years. B. Sachs extended our knowledge of the disease, a comparatively rare one, about 100 cases being reported in 1917 (Neville).

**Etiology.**—Among familial diseases it is unique in the limitation to one race—the Hebrew, and almost exclusively to the Polish branch. No other factor is known; syphilis is excluded. A dominant Mendelian character is present as 50 per cent. of the children are usually affected and 100 per cent. of the same sex. The cause is unknown. Sachs believes that the children are born with a nervous system so inadequate to meet the demands that the

cells, after performing their function for a few years or months, undergo complete degeneration. The disease comes into the category of Gowers' abiotrophies.

**Pathology.**—There is marked agenesis of the brain, with degenerative changes in the large pyramidal cells, and swelling of the dendrites. The degenerative changes are widely spread throughout the gray matter of the brain, the cord, and the spinal ganglia (Schaffer.) The retinal changes are due to a similar degeneration in the ganglion cells.

**Symptoms.**—Healthy at birth, and to the third or fourth month, the child then begins to be listless, moving the limbs very little, and, as time goes on, is not able to hold up the head or sit up. The muscles are flaccid, rarely spastic. There is progressive failure of vision and examination of the fundus shows a disappearance of the nerve cells of the retina and a cherry red spot in the region of the macula. Within a year a hitherto well-developed baby becomes marantic, completely blind, and death occurs as a rule before the end of the second year. The disease must be distinguished from the ordinary diplegias and paraplegias. It is not always easy as spasticity may be present, but the retinal changes are distinctive. There is no treatment.

A juvenile form occurring between the eighth and the twelfth year associated with blindness, but no macular changes, has been reported, and not in the Hebrew race. It is doubtful whether this is the same disease. Related to the Tay-Sachs' disease is the remarkable familial macular degeneration without dementia in which the disease starts about puberty.

## XII. MYOCLONIC EPILEPSY

This is a familial disorder, beginning in childhood with epilepsy, chiefly nocturnal, and followed by myoclonic attacks and progressive dementia. A majority of the cases have occurred in family groups and often in degenerate stock. Single cases may occur in normal families. Nothing is known of the causation; Lundborg suggests a thyroid origin. The changes found in the brain cortex have been those of chronic epilepsy and dementia.

**Symptoms.**—The onset, in childhood, is with nocturnal epilepsy, which in a year or two is followed by myoclonia, sometimes preceded by tremor. All the voluntary muscles are involved in short, quick, clonic spasms, which progressively increase in intensity. The child may at first have good and bad days, the latter following, as a rule, nights with severe epileptic seizures. The myoclonia grows worse and the patient falls into a state of dementia. The severe myoclonia attacks lead up to genuine epileptic seizures. There is a strong psychic feature which is intensified if the patient knows he is watched; bright lights, sounds, and handling the muscles have the same effect (Lundborg). The familial character and the nocturnal epilepsy separate it from the essential myoclonia of Friedreich. The treatment is that of epilepsy.

## D. DISEASES OF THE MENINGES

The spinal membranes may be affected separately but inflammatory processes often involve both. The process may be inflammatory (suppurative), hæmorrhagic or hypertrophic. Inflammation of the dura mater is

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termed pachymeningitis and of the pia mater leptomeningitis, but usually the comprehensive term meningitis is used. The symptoms and signs produced are due to involvement of the nerves and spinal cord rather than to changes in the meninges.

### I. DISEASES OF THE DURA MATER

#### (*Pachymeningitis*)

(1) **External Pachymeningitis.**—**CEREBRAL.**—Hæmorrhage often occurs as a result of fracture. Inflammation of the external layer of the dura is rare. Caries of the bone, either extension from middle-ear disease or due to syphilis, is the principal cause. In the syphilitic cases there may be a great thickening of the inner table and a large collection of pus between the dura and the bone.

Occasionally the pus is infiltrated between the two layers of the dura mater or may extend through and cause a dura-arachnitis.

The *symptoms* are indefinite. In acute cases signs of irritation are present. In the syphilitic cases there may be a small sinus communicating with the exterior. Compression symptoms may occur with or without paralysis.

**SPINAL.**—An acute form may occur in syphilitic affections of the bones, with retro-pharyngeal abscess, in tumors, and in aneurism. The symptoms are those of irritation of the nerve roots involved and compression of the cord. The chronic form is more common, and is a constant accompaniment of tuberculous caries of the spine. The internal surface of the dura may be smooth, while the external is rough and covered with caseous masses. The entire dura may be surrounded, or the process may be confined to the ventral surface.

(2) **Internal Pachymeningitis.**—This occurs in four forms: (1) Pseudo-membranous, (2) purulent, (3) hypertrophic and (4) hæmorrhagic. Pseudo-membranous inflammation of the lining membrane of the dura is not usually recognizable, but an example of it came under observation as a secondary process in pneumonia. Purulent pachymeningitis may follow an injury, but is more commonly the result of extension from inflammation of the pia. It is remarkable how rarely pus is found between the dura and arachnoid membranes. The hypertrophic form is usually syphilitic; the dura mater may be greatly thickened and involve the nerve roots and cord.

**Hæmorrhagic Internal Pachymeningitis.**—**CEREBRAL FORM.**—This remarkable condition, first described by Virchow, is very rare in general medical practice. During ten years no case came to autopsy at the Montreal General Hospital but in the post-mortem room of the Philadelphia Hospital, which received material from a large almshouse and asylum, the cases were not uncommon. The frequency in asylum work may be gathered from the fact that in 1,185 post mortems at the Government Hospital for the Insane, Washington, there were 197 cases with "a true neo-membrane of internal pachymeningitis" (Blackburn). Of these cases, 45 were chronic dementia, 37 were general paresis, 30 senile dementia, 28 chronic mania, 28 chronic melancholia, 22 chronic epileptic insanity, 6 acute mania, and 1 case imbecility. Forty-two were in persons over seventy years of age.

It has been found in profound anæmia and diseases of the blood-vessels, and has followed the acute fevers—typhoid fever in a child (Barker). It may occur in badly nourished cachectic children (Herter).

**Pathology.**—Virchow's view that the delicate vascular membrane precedes the hæmorrhage is undoubtedly correct. Practically we see one of three conditions: (a) subdural vascular membranes, often of extreme delicacy; (b) simple subdural hæmorrhage; (c) a combination of the two, vascular membrane and blood clot. The vascular membrane may exist without a trace of hæmorrhage—simply a fibrous sheet of varying thickness, permeated with large vessels, which may form beautiful arborescent tufts. On the other hand, there are instances in which the subdural hæmorrhage is found alone, but it is possible that in some of these the hæmorrhage may have destroyed all trace of the vascular membrane. In some cases a series of laminated clots are found, forming a layer from 3 to 5 mm. in thickness. Cysts may occur within this membrane. The source of the hæmorrhage is probably the dural vessels. Some hold that the bleeding comes from the vessels of the pia mater, but in the early stage there is no evidence of this; on the other hand, the highly vascular subdural membrane may be seen covered with the thinnest possible sheeting of clot, which has evidently come from the dura. The subdural hæmorrhage is usually associated with atrophy of the convolutions, and it is held that this is one reason why it is so common in the insane, especially in general paresis and dementia senilis. It occurs also in various cachectic conditions in which cerebral wasting is common. König found in 135 cases of hæmorrhagic pachymeningitis that 23 per cent. accompanied tuberculosis.

The **symptoms** are indefinite, or there may be none at all, especially when the hæmorrhages are small or have occurred very gradually, and the diagnosis can not be made with certainty. Headache has been a prominent symptom in some cases, and when the condition exists on one side there may be hemiplegia. The most helpful signs for diagnosis, indicating that the hæmorrhage in an apoplectic attack is *meningeal*, are (1) those referable to increased intracranial pressure, (slowing and irregularity of the pulse, vomiting, coma, contracted pupils, reacting to light slowly or not at all) and (2) paresis and paralysis, gradually increasing in extent, accompanied by symptoms which point to a *cortical* origin. Extensive bilateral disease may, however, exist without any symptoms whatever.

The *spinal fluid* may be bloody but this is not always the case. It is not a little curious that coma may come on and be the chief feature when anatomically the condition is a laminated hæmatoma evidently of long standing.

**SPINAL FORM.**—The spinal *internal pachymeningitis*, described by Charcot and Joffroy, involves chiefly the cervical region (*P. cervicalis hypertrophica*). The space between the cord and the dura is occupied by a firm, concentrically arranged, fibrinous structure, which arises within, not outside, the dura mater. It is anatomically identical with the cerebral internal hæmorrhagic pachymeningitis. The *etiology* is unknown; syphilis has existed in a few cases. The cord is usually compressed; the central canal may be dilated—*hydro-myelus*—and there are secondary degenerations. The *nerve roots* are involved and are damaged and compressed. The extent is variable. It may be limited

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to one segment, but more commonly involves a considerable portion of the cervical enlargement. Some cases present characteristic *symptoms*. There are intense neuralgic pains in the course of the nerves whose roots are involved. They are chiefly in the arms and in the cervical region, and vary greatly in intensity. There may be hyperæsthesia with numbness and tingling; atrophic changes may develop, and there may be areas of anæsthesia. Gradually motor disturbances appear; the arms become weak and the muscles atrophied, particularly in certain groups, as the flexors of the hand. The extensors remain intact, so that the condition of claw-hand is gradually produced. The grade of atrophy depends much upon the extent of involvement of the cervical nerve roots, and in many cases the atrophy of the muscles of the shoulders and arms becomes extreme. The condition is one of cervical paraplegia, with contractures, flexion of the wrist, and typical *main en griffe*. Usually before the arms are greatly atrophied there are the symptoms of what French writers term the second stage—involvement of the lower extremities and the gradual production of a spastic paraplegia, due to secondary changes in the cord.

The disease runs a chronic course, lasting, perhaps, two or more years. In a few instances recovery has taken place. The disease is to be distinguished from amyotrophic lateral sclerosis, syringomyelia, and tumors. From the first it is separated by the marked severity of the initial pains in the neck and arms; from the second by the absence of the sensory changes characteristic of syringomyelia. From certain tumors it is very difficult to distinguish; in fact, the fibrinous layers form a tumor around the cord.

The condition known as *hæmatoma* of the dura mater may occur at any part of the cord, or, in its slow, progressive form—pachymeningitis hæmorrhagica interna—may be limited to the cervical region. It is sometimes extensive, and may coexist with a similar condition of the cerebral dura. Cysts may occur filled with hæmorrhagic contents. The *treatment* of these conditions depends upon the cause. With injury or caries operation may be indicated. If syphilis is responsible active treatment should be given.

## II. DISEASES OF THE PIA MATER

### (*Acute Cerebro-spinal Leptomeningitis*)

**Etiology.**—Under cerebro-spinal fever and tuberculosis the two most important forms of meningitis have been described. Other conditions with which meningitis is associated are: (1) *The acute fevers*, more particularly pneumonia, erysipelas, and septicæmia; less frequently small-pox, typhoid fever, scarlet fever, measles, influenza, etc. (2) *Injury or disease of the bones of the skull*. In this group by far the most frequent cause is necrosis of the petrous portion of the temporal bone in chronic otitis. (3) *Extension from disease of the nose*. Meningitis has followed perforation of the skull in sounding the frontal sinuses, suppurative disease of these sinuses, and necroses of the cribriform plate. (4) As a *terminal infection* in chronic nephritis, arterio-sclerosis, heart disease, and the wasting diseases of children.

The following etiological table of the chief acute forms of meningitis may be useful to the student:

ACUTE LEPTOMENINGITIS.	Primary.	1. Of cerebro-spinal fever.	(a) Sporadic. } Meningococcus. (b) Epidemic. }	
		2. Pneumococcic.	Meninges involved alone or in a general pneumococcus or streptococcus infection.	Pneumococcus.
		3. Streptococcic.		Streptococcus.
	Secondary.	1. Tuberculous		Bacillus tuberculosis.
		2. Pneumococcic.	(a) Secondary to pneumonia, endocarditis, etc. (b) Secondary to disease or injury of cranium or its fossæ.	Pneumococcus.
		3. Pyogenic.	(a) Following local disease of cranium or a local infection elsewhere. (b) Terminal infection in various chronic maladies.	Various forms of staphylococci and streptococci.
		4. Miscellaneous acute infections.	In typhoid fever, influenza, diphtheria, gonorrhœa, anthrax, actinomycosis, and other acute diseases.	Typhoid bacillus, influenza bacillus, diphtheria bacillus, gonococcus, etc.

**Morbid Anatomy.**—The basal or cortical meninges may be chiefly attacked. The degree of involvement of the spinal meninges varies. In the form associated with pneumonia and ulcerative endocarditis the disease is bilateral and usually limited to the cortex. In extension from disease of the ear it is often unilateral and may be accompanied with abscess or with thrombosis of the sinuses. In the non-tuberculous form in children, in the meningitis of chronic nephritis, and in cachectic conditions the base is usually involved. In the cases secondary to pneumonia the effusion beneath the arachnoid may be very thick and purulent, completely hiding the convolutions. The ventricles also may be involved, though in these simple forms they rarely present the distention and softening so frequent in tuberculous meningitis.

**Symptoms.**—Cortical meningitis is not to be recognized by any symptoms or set of symptoms from a condition which may be produced by the toxins of many of the specific fevers. In the cases of so-called cerebral pneumonia, unless the base is involved and the nerves affected, the disease is unrecognizable since identical symptoms may be produced by intense engorgement of the meninges. In typhoid fever, in which meningitis is very rare, the twitchings, spasms, and retraction of the neck are almost invariably associated with toxæmia or meningism, not with true meningitis.

A knowledge of the *etiology* gives a very important clue. Thus, in middle-age disease the development of high fever, delirium, vomiting, convulsions, and retraction of the head and neck is extremely suggestive of meningitis or abscess. *Headache*, which may be severe and continuous, is the most common symptom. While the patient remains conscious this is usually the chief complaint, and even when semicomatose he may continue to groan and place his hand on his head. In the fevers, particularly in pneumonia, there may be no complaint of headache. *Delirium* is frequently early, and is most marked when the fever is high. *Photophobia* is often present. *Convulsions* are less common in simple than in tuberculous meningitis. In the simple meningitis of children they may occur. Epileptiform attacks which come and go are highly characteristic of direct irritation of the cortex. Rigidity and spasm or twitchings of the muscles are more common. Stiffness and retraction of the muscles of the neck are important signs; but they are not constant, and occur most often when the inflammation is extensive on the meninges

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of the cervical cord. There may be trismus, gritting of the teeth, or spastic contraction of the abdominal muscles. *Vomiting* is a common symptom in the early stages, particularly in basilar meningitis. Constipation is usually present. In the late stages the urine and faeces may be passed involuntarily. Optic neuritis is rare in the meningitis of the cortex, but is not uncommon when the base is involved. Marked *hyperæsthesia* is common.

Important signs are due to lesions of the *nerves* at the base. Strabismus or ptosis may occur. The facial nerve may be involved, producing slight paralysis, or there may be damage to the fifth nerve, producing anæsthesia, and, if the Gasserian ganglion is affected, trophic changes in the cornea. The pupils are at first contracted, subsequently dilated, and perhaps unequal. The reflexes in the extremities are often accentuated at the beginning of the disease; later they are diminished or entirely abolished. Herpes is common, particularly in the epidemic form.

*Fever* is present, moderate in grade, rarely rising above 103°. In the non-tuberculous meningitis of debilitated children and in nephritis there may be little or no fever. The pulse may be increased in frequency at first, though this is unusual. One of the striking features is the slowness of the pulse in relation to the temperature, even in the early stages. Subsequently it may be irregular and still slower. Very rapid emaciation often occurs, *Kernig's sign* has been described under cerebro-spinal fever. There may be a concomitant reflex of one leg when passive flexion is made of the other or when the neck is bent forward there is flexion of the legs both at the knees and hips or of all four extremities (Brudzinski's sign). *Lumbar puncture* is exceedingly valuable for diagnosis. The sugar in the spinal fluid is reduced or absent. A turbid fluid usually indicates an acute non-tuberculous meningitis. At first the fluid may be only opalescent. A close relationship exists between the severity of the symptoms, the height of the pyrexia, and the degree of turbidity (Connal). As a rule a preponderance of polynuclear leucocytes is present with the meningococcus or the pyogenic organisms; a mononuclear exudate occurs in tuberculosis or polio-myelitis. In tuberculous meningitis the fluid is usually clear; in only one of 69 cases was it opalescent (Connal).

**Diagnosis.**—The discussion under cerebro-spinal fever applies. One should decide first if meningitis is present, and next the etiology. In both a study of the cerebro-spinal fluid is the most important aid.

**Treatment.**—The patient should be kept as quiet as possible and causes of irritation removed. An ice-bag should be applied to the head. Sedatives should be given if the patient is conscious and suffering. The bowels should be freely moved. Warm baths should be given every three hours. Large doses of iodide of potassium and mercury are recommended by some authors. Hexamine in doses of 60 grains (4 gm.) daily may be tried, as Crowe has shown that it is excreted in the cerebro-spinal fluid and inhibits the growth of organisms in the meninges. When the causal organism is determined serum treatment may be used if available. The meningococcus form has been discussed. With a meningitis due to Type I pneumococcus the serum should be used freely both intraspinally and intravenously. In meningitis due to the influenza bacillus the use of serum is indicated. In the streptococcus form it is not likely to be of value but should be tried.

Lumbar puncture, as a therapeutic measure, is of great value, relieving the headache and sometimes reducing the fever.

**MENINGISM.**—Sometimes spoken of as the syndrome of Dupré, this is a condition in which there are symptoms of meningitis, but post mortem the characteristic pathological changes are not present. It is practically the condition described formerly as meningeal irritation, and is seen most frequently in the acute fevers of children, particularly in pneumonia and typhoid fever, sometimes in alcoholism and in middle-ear disease. Lumbar puncture usually gives a large amount of clear, sterile fluid, under increased pressure and sometimes showing a slight increase in the number of cells. While many disturbances are due to the fluid being under increased pressure, subnormal pressure may be responsible for symptoms. Headache, dizziness, and possibly dullness (semi-comatose) may be due to deficiency of cerebro-spinal fluid. Similar features may be seen after lumbar puncture. The condition is corrected, usually rapidly, by subcutaneous saline injections.

**CHRONIC LEPTOMENINGITIS.**—This is rarely seen apart from syphilis or tuberculosis, in which the meningitis is associated with the growth of the granulomata in the meninges and about the vessels. The symptoms in such cases are extremely variable, depending entirely upon the situation of the growth. The meningococcus meningitis may run a very chronic course, but of all forms the posterior basic may be the most protracted, as cases have been described with a duration of a year or more. Quincke's *meningitis serosa* is considered with hydrocephalus.

## E. MENINGO-MYELO-ENCEPHALITIS

### I. ACUTE POLIO-MYELITIS

#### (Heine-Medin Disease)

**Definition.**—An acute infection characterized anatomically by widespread lesions of the nervous system, with special localization in many of the cases in the anterior horns of the gray matter in the spinal cord—hence the common name, *polio-myelitis anterior*.

**History.**—In 1840 von Heine separated this type from other forms of paralysis and in 1887 Medin called attention to its occurrence in widespread epidemics, which have been specially studied in Sweden by Wickham, Harnitz, and others. Serious outbreaks have occurred in many parts of the United States and Canada. The incidence of the disease has increased in Great Britain and Europe; in Sweden and Norway and parts of Austria the disease has assumed epidemic proportions. In New York City in 1907-8 there were about 2,000 cases, with a mortality of 6 to 7 per cent.; in 1916 in the U. S. registration area there were 7,130 deaths and 769 in 1920.

**Etiology.**—In its epidemic behavior the disease resembles closely cerebro-spinal fever. Sporadic cases occur in all communities and under at present unknown conditions increase at times to epidemic proportions. It prevails especially in the late summer and autumn.

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*Age* is an important element; a majority of cases occur in young children. The more prevalent the epidemic the greater the proportion of young adults attacked. Males and females are about equally attacked.

The degree of infectiousness from person to person is slight, and in this the disease resembles cerebro-spinal fever and pneumonia.

The *organism* was isolated by Flexner and his co-workers. The colonies consist of globular bodies averaging 0.15 to 0.3 micron in size. Monkeys inoculated with the twentieth generation of the culture developed typical experimental polio-myelitis. The infective agent is present in the brain and spinal cord, in the naso-pharyngeal secretions and in the blood. The disease is inoculable into monkeys and may be transmitted from one animal to another. It has been transmitted also by intracerebral injection of an emulsion made from flies which had fed on the spinal cord of a monkey dead of the disease. An important point is that the virus passes from the central nervous system in the monkey to the nasal mucosa and vice versa, and the application of the virus to this part is a ready means of inoculation. It has also been found in the tonsils and pharyngeal mucosa of children. The path of invasion is apparently by the upper respiratory tract.

So far as we know, the disease is transmitted either directly by contact or by the intervention of carriers. The distribution is more independent of sanitary conditions than in the common children's diseases.

**Morbid Anatomy.**—The lesions are widespread in the nervous system. It is not an affection limited to the anterior horns of the gray matter of the spinal cord, but a widespread polio-myelo-encephalo-meningitis.

Swelling of the spleen and a marked general hyperplasia of the lymphoid apparatus are found. The cerebro-spinal fluid is usually increased but clear. The pia mater is hyperæmic and moist, but without exudate. Cases in which the cerebral symptoms have been pronounced show swelling and flattening of the convolutions, with hyperæmia of the gray matter and here and there small hæmorrhages. The changes in the spinal cord are very characteristic. The meninges are moist, the pia is hyperæmic, sometimes with small capillary hæmorrhages. On section the cut surface bulges, the gray matter is hyperæmic, appearing as a reddened H, or the redness is limited to the anterior horns, which may show spots of hæmorrhage. These changes may be localized to the swellings of the cord or extend throughout its entire extent. Microscopically there is small-celled infiltration about the vessels of the meninges, most marked in the lumbar and cervical swellings. The infiltration extends into the fissures of the cord and follows the blood-vessels. The amount of meningeal implication is much more intense than is indicated macroscopically. In the cord itself the smaller blood-vessels are distended, hæmorrhages occur in the gray matter, there is marked perivascular infiltration, chiefly of lymphocytes, which collect about the vessels, forming definite foci. Sometimes the majority of the cells are polynuclear leucocytes. The ganglion cells, usually those of the anterior horns, degenerate and gradually disappear, changes probably secondary to the acute vascular alterations and toxæmia. Hyperæmia, œdema and infiltration are marked. After the inflammation subsides, the nerve cells may be destroyed, or only damaged so that some recovery is possible. Sclerotic changes follow in the affected areas. The muscles innervated from the damaged areas atrophy depending on the amount

of destruction of the nerve cells. In the fatal cases there are changes in the medulla and pons of much the same nature, but the ganglion cells rarely show such widespread destruction.

**Symptoms.**—The incubation period is from 3 to 10 days. In the pre-paralytic stage naso-pharyngeal symptoms are common. F. R. Fraser notes among the pre-paralytic symptoms, fever, drowsiness or heaviness, irritability, twitchings and jerkings, and gastro-intestinal symptoms. In 72 of 90 cases there was stiffness of the neck and back; general tenderness (hyperæsthesia) on handling is not at all uncommon. Sometimes a child who has gone to bed well awakens in the morning with paralysis and slight fever. Prodromal symptoms are more common in the epidemic form. The paralysis is of the flaccid form, with loss of the deep reflexes in affected areas and subsequent atrophy. The paralysis is usually most widespread at the onset and gradually lessens. The temporary paralysis is due to inflammatory and toxic changes while the permanent paralysis depends on destruction of nerve cells. In the early stages there is usually a well-marked polynuclear leucocytosis.

There are a number of forms, of which the most important are:

(a) **ABORTIVE FORM.**—There are cases of illness with the general symptoms of infection, and indications of cerebro-spinal irritation, but without any motor disturbances. The symptoms pass away and the nature of the trouble remains doubtful, nor would suspicion be aroused were it not for the existence of other cases. Anderson and Frost have shown the presence of specific immune bodies in the blood of these cases.

(b) **COMMON POLIO-MYELITIC OR SPINAL FORM.**—There is paresis before the paralysis or the paralysis is abrupt in its onset and reaches its maximum in a very short time, showing the irregularity and lack of symmetry which is characteristic. The legs are involved much more often than the arms. Paralysis of the trunk muscles occurs often. One or both arms may be affected, or one arm and one leg, or both legs. In the arm the paralysis is rarely complete, the upper-arm muscles may be most affected or the lower-arm group muscles acting functionally together, with centres near each other in the spinal cord, are paralyzed together. The trunk muscles may show paralysis. Careful examination usually shows some degree of weakness to be more widespread than appears at first sight. Disturbances of sensation are common, especially tenderness of the muscles. In this type the bladder and rectum are rarely involved.

(c) **PROGRESSIVE ASCENDING FORM.**—A certain number of cases, particularly in epidemics, run a course similar to Landry's paralysis, with which, no doubt, some of them have been confounded. The disease begins in the legs with the usual initial symptoms, the paralysis extends upward, involving the arms and the trunk, and death may occur with bulbar symptoms from the third to the fifth day. In the Swedish epidemic of 1905 of the 159 cases which died within the first two weeks, 45 presented this form.

(d) **BULBAR FORM.**—In this the cranial nerves are involved and a wide variety of lesions may result. There may be difficulty in breathing and swallowing with paralysis of the ocular, facial, lingual or pharyngeal muscles. The onset may be sudden. The patient has fever, and the local picture depends upon the extent and distribution of the lesions in the medulla and pons. A fatal result is common.

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(e) **MENINGITIC FORM.**—This is important, as the cases simulate closely and are apt to be mistaken for cerebro-spinal fever. The picture is one of an acute meningitis—headache, pain and stiffness in the neck, vomiting, pain and rigidity in the back, drowsiness and unconsciousness. The disease may begin with the paralytic features and subsequently show the meningeal complications. Convulsions and Kernig's sign may be present. A serious difficulty is that the two diseases may prevail together, and only the careful examination of the cerebro-spinal fluid may give a differential diagnosis.

(f) **CEREBRAL FORM.**—Here the picture is that which we have learned to recognize as the acute encephalitis or polio-encephalitis of children, a description of which we owe to von Strümpell. The disease sets in suddenly, with fever, vomiting and convulsions, followed by paralysis of one side of the body or one limb. Many of the patients die, others recover and present the usual after-picture of the cerebral hemiplegia of children. A large proportion of these cases probably results from this form of acute polio-myelo-encephalitis. Athetosis and epilepsy may be sequels.

(g) **CEREBELLAR FORM.**—In this there is marked ataxia with retraction of the head and rigidity of the neck, but not necessarily nystagmus. Acute vestibulitis may give a similar picture for a few days, but complete bilateral deafness follows.

(h) **POLYNEURITIC FORM.**—It is one of the features of the epidemic form that the patients complain much more of pain. This is particularly the case in a form which simulates polyneuritis. There is loss of the tendon reflexes and disturbance of sensation. There is pain in the affected limbs, particularly on movement, with tenderness on pressure along the nerves and on pressing the muscles; the paralysis may extend like neuritis, involving chiefly the peripheral extensor muscle groups, and be followed by rapid wasting.

(i) **TRANSVERSE MYELITIC FORM.**—Following slight fever and indisposition, the features may be those of a transverse myelitis, a complete flaccid paraplegia. Of two cases of this type in young adults, in one recovery was complete, and in the other with a very small amount of residual paralysis.

*Anomalous forms* are common during an epidemic. The muscles of respiration may be involved early, the diaphragm alone may be paralyzed, or the intercostals or the muscles of the palate and pharynx. Involvement of the facial muscles, usually a slight weakness, may be present, but in 5 out of 90 cases studied by F. R. Fraser the facial muscles alone were involved. In one instance ptosis was the only paralytic symptom on admission. Remarkable forms occur quite unlike the classical picture. In one case there was paralysis of one side of the soft palate with slight fever; the serum of this patient protected a monkey from intra-cerebral injection of the polio-myelitic virus. There may be slight fever with general spasticity of the muscles and tremor or rigidity of the muscles with coma.

*Spinal Fluid.*—This usually shows increase both in amount and pressure; it may be clear or slightly hazy. There is an increase in the number of cells, which may be from 15 or 20 up to 1200 per c. mm. The largest number are usually mononuclears; occasionally a larger number of polynuclears is found early. The albumin and globulin are usually increased. Fehling's solution is generally reduced as promptly as by the normal fluid. The Wassermann reaction may be the only means of diagnosing the condition from syphilis.

**Course.**—Usually within a few days the extent of paralysis lessens and the improvement is rapid for a short time; after this it is slow but may continue for two or three months. The residual paralysis is usually less than seemed probable at first. Return of the reflexes is a hopeful sign. The atrophy becomes evident in a few weeks from the onset of the attack. The affected limbs show less development as the patient grows older, and the deformity is usually most marked in the leg. The reaction of degeneration is present in the atrophied muscles. Early in the course the muscles lose the faradic response.

**Diagnosis.**—In the early stages before paralysis appears, this is difficult; the occurrence of an epidemic and the findings in the cerebro-spinal fluid are suggestive. When paralysis occurs in the ordinary spinal cases there is rarely any difficulty. An important point to remember is that during epidemics the disease presents an extraordinary number of forms. Some cases run a course like an acute infection, others have the picture of Landry's paralysis, in others again meningeal symptoms predominate, or there may be hyperæsthesia and pain, with the picture of a polyneuritis.

It is not improbable that some obscure cases of meningitis are instances of polio-myelitis. The same may be said of the acute encephalitis in children causing hemiplegia. The complexity of the symptoms makes the diagnosis difficult, so that the study of the spinal fluid is important.

The diagnosis from peripheral neuritis may be very difficult; in both the paralysis is of the legs, with wasting, loss of reflexes, and the bladder and rectum may be involved. Loss of the vibrating sensation tested with a large tuning fork is more common in peripheral neuritis, and later the electrical changes and the reaction of degeneration may be distinctive.

**Prognosis.**—The mortality varies greatly in different epidemics. It was 27 per cent. in New York City in 1916. The fatal cases are usually of the ascending, bulbar and meningeal types. As regards the muscles, complete loss of response to faradism means severe atrophy. If it is never completely lost the outlook is good and even extensive paralysis may disappear. The prognosis for the paralysis is not easy to determine. Formerly, we thought it almost the rule that residual paralysis would remain if any large number of muscle groups were involved, but cases of very severe and widespread involvement may recover gradually and completely.

**Prophylaxis.**—The disease has been made notifiable. The patient should be isolated, the discharges and articles used by patients and nurses carefully disinfected, and special care taken of the nasal and pharyngeal discharges. It does not seem necessary to enforce a quarantine against those who come into relation with the patients, but the throats and noses of such persons should be disinfected with a menthol spray. There is some warrant for the administration of prophylactic doses of hexamine.

**Treatment.**—The patient should be at rest and given the usual treatment during the febrile stage. Salicylates can be given for pain and discomfort with codeine or morphine if necessary. Hexamine should be given in doses of gr. 5 to 15 (0.3-1 gm.). Lumbar puncture should be done at once and repeated in twelve hours if much fluid is obtained. The puncture should be repeated daily for three or four days. The spinal fluid should be drained freely. If there is respiratory difficulty atropine should be given freely, and

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with respiratory failure artificial respiration should be carried on. In the meningeal form hot packs are useful. Blood serum from one who has recovered from the disease, if available, is useful, in the early stages intraspinally and intravenously. Intraspinally less should be given than the amount of fluid removed. Intravenously 40 c. c. can be given to children up to two years and up to 80 c. c. to children ten years of age. Serum tinged with hæmoglobin should be given intramuscularly. Rosenow advises immune horse serum prepared by repeated injections of a pleomorphic streptococcus given intravenously and intramuscularly.

The muscles involved should be given as complete rest as possible. The affected limbs should be wrapped in cotton wool and placed in the position which gives the greatest physiological rest, so that they are not stretched by opposing muscles. Voluntary movements should not be allowed for some weeks. Educational movements and the use of apparatus demand the greatest care not to fatigue the muscles. Careful massage and passive movements may be practised. Electricity may be used and perhaps has a value in keeping up the nutrition of the muscles. The faradic current should be employed if there is response, if not, the galvanic. Fatigue is harmful and should be guarded against for many months.

The muscle itself as a factor has been emphasized by William MacKenzie of Melbourne as biologically it is all important in treatment. The disease really destroys muscle adjustments, and one of the first things to do is to place the muscle at physiological rest in the zero position, in which it is itself relaxed, and both its own action, and that of its opponent prevented. Massage, he urges, should not be given too early, until, for example, the patient can elevate the upper limb when sitting up, and the heel when lying on the back. Persistent gradual re-education of the muscles yields remarkable results. Passive movements may be used and with toys a child may be encouraged to use the muscles of any group which still act. The treatment of residual deformities is a question of orthopaedic surgery.

### II. EPIDEMIC ENCEPHALITIS

(*Encephalitis lethargica; Epidemic Stupor; Epidemic Polio-encephalitis; Infective Encephalitis*)

**Definition.**—An infectious disease, with protean manifestations, chiefly in the central nervous system, characterized by lethargy, paralysis of the cranial nerves (usually the third), and in some cases, spinal and neuritic features.

**History.**—There are records of outbreaks suggesting this disease in 1712 in Germany and in 1890 in parts of southern Europe (to which the name Nona was given). Cases occurred in Austria and France in 1916 and in England in the spring of 1918. The disease was widely spread, only a few cases occurring in each locality, sometimes two or three in the same house. The disease was recognized in the United States about the end of 1918. In England and Wales there were 1470 cases in 1921 and in France it is estimated that there were 10,000 cases up to 1920.

**Etiology.**—Males and females are attacked in about equal numbers. In striking contrast to polio-myelitis it is rare in young children and most com-

mon in young adults; cases in patients over the age of fifty are not uncommon. While the disease has resemblances to polio-myelitis, they are distinct entities. There have been very few epidemics in institutions. The nature of the virus has not been determined; the disease apparently has been successfully transmitted to monkeys.

**Pathology.**—The brain may have a pink or rose color and is hyperæmic. Hæmorrhages in the meninges and in the region of the basal ganglia may be found. The situation of the lesions varies markedly and while any part may be affected the mid-brain, pons, and basal ganglia are particularly involved. Lesions occur in the cortex, cerebellum and spinal cord. Histologically there is vascular congestion with marked lymphocytic infiltration about the vessels and cellular infiltration of the nerve tissues with œdema. The areas of extra-vascular infiltration may form visible foci. The lesions occur in nodular and diffuse forms. The nerve cells show degeneration which may be very local and irregularly distributed. Thrombosis and necrosis are rare. The cranial nerve paralyses are due to involvement of the nuclei and congestion and infiltration have been described in the roots of the cranial and spinal nerves. The gray matter at the base of the brain is particularly involved. The meninges may show inflammation. Altogether the anatomical lesions are like those found in rabies and sleeping sickness. The spinal cord lesions are usually slight. The lethargy may be toxic but is possibly mechanical due to interruption of stimuli in the thalamus, which is frequently involved. The later changes involve (1) the vessels with hyaline and calcareous degeneration of the media and adventitia especially and particularly in certain areas, especially the basal ganglia, (2) hydrocephalus, chronic or intermittent, from imperfect drainage of the ventricles, and (3) meningeal thickening.

**Clinical Features.**—These comprise the features of an infectious disease with the special manifestations due to the lesions in the nervous system. When the marked possibilities of variation in the situation of these lesions are considered, the diverse results and the polymorphous character of the symptoms and signs can be understood. It is natural that different forms are described in consequence. Lethargy is not characteristic of all; there is a very definite myoclonic form. In all the involvement of the cranial nerves is important.

The incubation period is variable and uncertain. Prodromal symptoms range from a few hours to a week, and are chiefly headache, lethargy, stiffness in the back, diffuse pains, and catarrhal features. The general features are of an acute infection. There is usually fever at the onset which, as a rule, is not high and does not last long but in some cases persists for weeks. It may disappear and recur. Headache, general malaise, and gastro-intestinal disturbance are common. There may be vomiting and constipation is generally marked. Local pains may be marked, probably due to involvement of the nerve roots. A number of local manifestations may occur, such as sweating, salivation, dysphagia, hiccup and retention of urine. Convulsions are rare. The signs due to involvement of the nervous system are of the greatest variety and all kinds of variations and combinations are seen.

*Lethargy*, present in 80 per cent. of the cases, comes on as a rule gradually, occasionally very suddenly, and is generally not more than a stupor and heavi-

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ness, from which the patient can easily be roused, the so-called "anergic apathy," but in others it is much deeper, passing into coma. As the disease progresses, the patient presents a dull apathetic look. The wrinkles are smoothed, the muscles of the face may be moved with <sup>given</sup> difficulty, or there may be definite bilateral facial paralysis. The arms are flexed, and catalepsy is not uncommon. When roused the patient may answer simple questions intelligently. In some cases the picture is very different. Active delirium or violent mania may be present. Persistent insomnia may <sup>be</sup> present, about which the patient may not specially complain. There may be <sup>at</sup> a condition of euphoria. Some patients talk incessantly; others <sup>are</sup> irritable and restless. In some cases the drowsiness combined with ataxia suggests alcoholic intoxication. The speech may be blurred and difficult; this depends on the degree of involvement of the facial muscles. Tremors, twitchings, and marked choreiform movements may occur, and persist long into convalescence.

*Cranial Nerves.*—These are nearly always involved at some time and to some degree even if transient and slight. A history of double vision is common. The pupils may show inequality, irregularity, and a sluggish reaction to light, or the Argyll-Robertson sign. Ptosis is common, often bilateral and sometimes overlooked on account of the lethargy. Paralysis of the eye muscles is frequent, it may be so slight as to be made out with difficulty or diplopia and strabismus may be marked. The third nerve is most frequently involved, the sixth frequently. Nystagmus is common. The seventh nerve is frequently affected; the process is usually unilateral. Involvement of the other cranial nerves is not as common.

*Disturbance of Movement.*—There may be the greatest variety of disturbance of muscular movement. Fibrillary tremors and twitching are common. Involuntary movements may involve a small area; thus there may be rhythmic movements of small extent about the forehead or face. They are often seen in the lower thorax or abdomen; one rectus muscle may show it. Of special interest is the *myoclonic form* in which there are widespread rhythmic contractions. The onset may be with severe pain. The contractions may be so marked that the patient is thrown out of bed. The muscles of the abdomen and legs are specially involved. It is probable that some of the cases of prolonged hiccough are due to this. In other cases the movements are athetoid or choreiform, or resemble those of paralysis agitans. Slowness of movement with rigidity is common, giving the mask-like appearance of the face and immobility of the body. Paralysis of all kinds may occur; spasticity with increased deep reflexes and an extensor plantar response may be found.

*Sensory* disturbances are rare. There may be pain, particularly on pressure of the muscles, and there is sometimes hyperæsthesia. The reflexes may vary greatly from day to day; the knee-jerks may be abolished for a time. Sphincter features are sometimes present. Dysphagia has been recorded in a number of cases.

*Forms.*—For convenience various forms are described, much as in poliomyelitis, and it is evident that the possible combinations are many. (1) So-called "abortive" cases with mild or transient manifestations. These are probably common. (2) Cases with general features but no localizing signs. The latter may be very slight and of short duration but are rarely absent entirely.

(3) The *lethargic* form, usually with localizing signs. (4) *Psychical* forms in which delirium and mental features predominate; paralyzes or myoclonus may appear later. (5) *Labyrinthian* form with vertigo or loss of equilibrium. (6) *Cerebellar* form with a picture of acute cerebellar ataxia. (7) *Bulbar* form. (8) *Meningeal* form. Meningitis may be closely simulated and in case of hemorrhage, the spinal fluid may contain blood. (9) *Neuritic* form. (10) *Spinal* form. This may show paralyzes like polio-myelitis or a picture like myelitis. (11) *Myoclonic* form. This seems to have become more frequent of late in the United States. (12) There is a form much like *myasthenia gravis*. Some cases show features of multiple sclerosis. (14) A hemorrhagic form is described. Hemorrhages may occur in the nervous system, from the mucous surfaces, or in the skin.

*Respiratory* disturbances are common, spasmodic cough, transient attacks of dyspnoea, rapid or slow respiration, especially the former, periods of apnoea, respiratory ties, and sniffling.

Signs suggestive of *meningeal* involvement occur. The cerebro-spinal fluid is, as a rule, clear with 10 to 20 cells per c. mm. (rarely 100 cells), mostly mononuclears, the globulin little, if at all, increased. The reducing substance may be increased. The colloidal gold test gives a luetic curve.

The **course** is extremely variable and the disease may persist for months. In some cases recovery takes from months to two years. Relapses or exacerbations may occur. Of special importance are the *sequels* which occur in great variety and in many combinations. Among them are psychical changes, which may progress to dementia, alterations in disposition and conduct, headache, marked neurasthenic states and insomnia. Disturbances of the sympathetic system may be marked. Respiratory disturbances have been observed. Nocturnal excitement has been noted, especially in children. Organic changes may be shown by ptosis, pupillary changes, nystagmus, strabismus, facial asymmetry, difficulty in speech or swallowing, paralysis of an extremity, and pyramidal tract involvement. Tremors are common, of the tongue, facial muscles or extremities. A large group shows the picture of *paralysis agitans*, which may appear after a long interval, with an expressionless face and alteration in the attitude with tremors and disturbance of gait. The resulting tremors are variable; some are choreiform or athetoid. Tics and torsion spasm are seen. Diabetes insipidus and obesity have followed an attack.

**Diagnosis.**—Typical cases offer no difficulty. Special watch should be kept for the cranial paralysis which in mild cases may be of short duration. The following conditions deserve mention: (1) Polio-myelitis. The similarity may be marked but the spinal fluid usually shows more marked changes in this disease. (2) Psychoses characterized by stupor, lethargy or catalepsy. (3) Tuberculous meningitis in which the spinal fluid findings are not characteristic. (4) Acute syphilitic meningo-encephalitis and endarteritis. (5) Botulism. (6) Cerebral hemorrhage or thrombosis may be simulated by some cases of encephalitis. (7) Status epilepticus. (8) Uraemia. (9) Other forms of encephalitis. The diagnosis may be made by the sequels after an unrecognized attack.

**Prognosis.**—An acute onset with rapid development of severe toxæmia, marked delirium, high fever, coma and marked myoclonic features is of grave

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men. The mortality is probably about 20 to 25 per cent. in the acute stages. Sudden death occurs especially in the bulbar form. The ultimate prognosis is uncertain and the frequency of sequels renders the outlook for complete recovery always doubtful. The appearance of the features like paralysis agitans is serious. Improvement may be very slow but is sometimes continued for a long time.

**Treatment.**—The patient should be isolated and carefully nursed, care being taken to avoid bed-sores; nasal and rectal feeding may be required and special care should be taken to keep the mouth and throat clean. Special care is to be given to the bowels, which should be kept freely open. Retention of urine should be avoided. Lumbar puncture should be done at once and repeated daily for some days, the number of times being governed by the results. There is no specific treatment. Hexamine has been given in full doses. Sedatives may be required for headache or pain. The insomnia is resistant and full doses of hypnotics may be necessary. In convalescence, hot baths, massage and passive motion may be used for rigidity. As much encouragement as possible should be given. For the Parkinsonian syndrome hyoscine hydrobromide in doses up to gr. 1/50 (0.0013 gm.) three times a day may be useful. Pheno-barbital seems to be beneficial in some patients.

## F. MYELITIS

### I. ACUTE MYELITIS

**Etiology.**—Acute myelitis affecting the cord in a limited or extended portion—the gray matter chiefly, or the gray and white matter together, is met with: (a) As an independent affection probably due to infection, and leading to rapid loss of power with the symptoms of an acute ascending paralysis. Some of these cases are unusually widespread acute forms of poliomyelitis. There is also an acute hemorrhagic form with high fever (Burley), the relation of which to other forms is uncertain. (b) As a sequel of the infectious diseases, such as small-pox, typhus, measles, and gonorrhœa. (c) As a result of *traumatism*, either fracture of the spine or very severe muscular effort. Concussion without fracture may produce it, but this is rare. Acute myelitis scarcely ever follows railway accidents. (d) In diseases of the *bones* of the spine, either caries or cancer. This is a more common cause of localized acute transverse myelitis than of the diffuse affection. (e) In disease of the cord, such as *tumors* and *syphilis*; in the latter in association with gummata, in which case it is usually a late manifestation, or it may follow within a year of the primary affection. In the syphilitic form, the process may be secondary to vascular disease or to meningeal involvement. In cases of long standing marked sclerosis results, and in a transverse lesion there is ascending and descending secondary degeneration.

**Morbid Anatomy.**—In localized acute myelitis affecting white and gray matter, as met with after accident or an acute compression, the cord is swollen, the pia injected, the consistence greatly reduced, and on incising the membrane an almost diffuent material may escape. In less intense grades, on

section at the affected area, the distinction between the gray and white matter is lost, or is extremely indistinct. There are cases with the appearances of an acute hæmorrhagic myelitis.

**Symptoms.**—(a) ACUTE DIFFUSE MYELITIS.—This form is in the epidemic polio-myelitis, or occurs in connection with syphilis or one of the infectious diseases, or is seen in a typical manner in the extension from injuries or from tumor. The onset, though scarcely so abrupt as in hæmorrhage, may be sudden; a person may be attacked on the street and have difficulty in getting home. In some instances, the onset is preceded by pains in the legs or back, or a girdle sensation is present. It may be marked by chills, occasionally by convulsions; fever is usually present from the beginning—at first slight, but subsequently it may become high.

The *motor* functions are rapidly lost, sometimes as quickly as in Landry's ascending paralysis. The paraplegia may be complete, and, if the myelitis extends to the cervical region, there may be impairment of motion, and ultimately complete loss of power in the upper extremities as well. The sensation is lost, but there may at first be hyperæsthesia. The *reflexes* in the initial stage are increased, but in acute central myelitis, unless limited in extent to the thoracic and cervical regions, the reflexes are usually abolished. The rectum and bladder are paralyzed. Trophic disturbances are marked; the muscles waste rapidly; the skin is often congested, and there may be localized sweating. The temperature of the affected limbs may be lowered. Acute bed-sores may occur over the sacrum or on the heels, and sometimes a multiple arthritis is present. In these acute cases the general symptoms become greatly aggravated, the pulse is rapid, the tongue becomes dry; there is delirium, the fever increases, and may reach  $107^{\circ}$  or  $108^{\circ}$  F.

The *course* of the disease is variable. In very acute cases death follows in from five to ten days. The cases following the infectious diseases, particularly the fevers and sometimes syphilis, may run a milder course.

(b) ACUTE TRANSVERSE MYELITIS.—The symptoms naturally differ with the situation of the lesion.

(1) Acute transverse myelitis in the *thoracic region*, the most common situation, produces a very characteristic picture. The symptoms of onset are variable. There may be initial pains or numbness and tingling in the legs. The paralysis may set in quickly and become complete within a few days; but more commonly it is preceded for a day or two by sensations of pain, heaviness, and dragging in the legs. The paralysis of the lower limbs is usually complete, and if at the level, say, of the sixth thoracic vertebra, the abdominal muscles are involved. Sensation may be partially or completely lost. At the onset there may be numbness, tingling, or even hyperæsthesia in the legs. At the level of the lesion there is often a zone of hyperæsthesia. A girdle sensation may occur early, and when the lesion is in this situation it is usually felt between the ensiform and umbilical regions. The reflexes are variable. There may at first be abolition; subsequently, those which pass through the segments lower than the one affected may be exaggerated and the legs may take on a condition of spastic rigidity. It does not always happen, however, that the reflexes are increased here, for in a total transverse lesion of the cord they are usually entirely lost, as pointed out by Bastian. That this is not due to the preliminary shock is shown by the fact that the abolition may

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be permanent. The muscles become extremely flabby, waste, and lose their faradic excitability, and the sphincters lose their tone. The temperature of the paralyzed limbs is variable. It may at first rise, then fall and become subnormal. Lesions of the skin are not uncommon, and bed-sores are apt to form. There is at first retention of urine and subsequently spastic incontinence. If the lumbar centres are involved, there are vesical symptoms from the outset. The urine is alkaline and may rapidly become ammoniacal. The bowels are constipated and there is usually incontinence of feces.

(2) *Transverse Myelitis of the Cervical Region.*—If the lesion is at the level of the sixth or seventh cervical nerves, there is paralysis of the upper extremities, more or less complete, sometimes sparing the muscles of the shoulder. Gradually there is loss of sensation. The paralysis is usually complete below the point of lesion, but there are rare instances in which the arms only are affected, the so-called cervical paraplegia. In addition to the symptoms already mentioned there are several which are more characteristic of transverse myelitis in the cervical region, such as the occurrence of vomiting, hiccough, and slow pulse, which may sink to 20 or 30, pupillary changes—myosis—sometimes attacks of dysphagia, dyspnoea, or syncope.

The course of complete transverse myelitis depends upon its cause. Death may result from extension. Segments of the cord may be completely and permanently destroyed, in which case there is persistent paraplegia. The pyramidal fibres below the lesion undergo the secondary degeneration, and there is an ascending degeneration of the dorsal median columns. If the lower segments of the cord are involved the legs may remain flaccid. In some instances a transverse myelitis of the thoracic region involves the ventral horns above and below the lesion, producing flaccidity of the muscles, with wasting, fibrillary contractions, and the reaction of degeneration. More commonly, however, in the cases which last many months there is more or less rigidity of the muscles with spasm or persistent contraction of the flexors of the knee. The *prognosis* is always serious as regards complete recovery. The syphilitic cases usually make the best recovery if recognized early and properly treated.

The *diagnosis* of myelitis is rarely difficult. In common with the acute ascending paralysis of Landry, and with certain cases of multiple neuritis, it presents a rapid and progressive motor paralysis. From the former it is distinguished by more marked involvement of sensation, trophic disturbances, paralysis of bladder and rectum, rapid wasting, electrical changes, and fever. From acute cases of *multiple neuritis* it may be more difficult to distinguish, as the sensory features may be marked, though there is rarely, if ever, in multiple neuritis complete anaesthesia; the wasting, moreover, is more rapid in myelitis. The bladder and rectum are rarely involved—though in exceptional cases they may be—and, most important of all, the trophic changes, the development of bullae, bed-sores, etc., are not seen in multiple neuritis. An *etiological* diagnosis is important, especially for treatment. Syphilis should always be considered; the spinal fluid study is usually definite. The possibility of pressure from disease of the vertebrae or by tumor has to be excluded.

**Treatment.**—In the rapidly advancing form due either to a diffuse inflammation in the gray matter or to transverse myelitis, the important measures are scrupulous cleanliness, care and watchfulness in guarding

against bed-sores, and the avoidance of cystitis. In an acute onset in a healthy subject the spine may be cupped. Counter-irritation is of doubtful advantage. Chapman's ice bag is sometimes useful. No drugs have the slightest influence upon acute myelitis, except in subjects with well-marked syphilis, in which case mercury and potassium iodide should be given energetically. Mercury seems to be much more useful than asphenamine, which may be given in the intervals between courses of mercury. Tonic remedies, such as arsenic and strychnine, may be used in the later stages. When the muscles have wasted, massage is beneficial in maintaining their nutrition. The patient should make every effort to perform muscular movements himself and thus aid improvement. Electricity should not be used in the early stages. It is of no value in the transverse myelitis in the thoracic region with retention of the nutrition in the muscles of the leg.

## II. ACUTE ASCENDING (LANDRY'S) PARALYSIS

**Definition.**—An acute ascending flaccid paralysis beginning in the legs and spreading upwards, without loss of reflexes or disturbance of sensation. The termination is in complete recovery or death, in which case no gross nervous system lesions are found.

**Etiology and Pathology.**—The disease occurs most commonly in males between the twentieth and thirtieth years. It has followed the specific fevers and various organisms have been isolated. There is a form of the epidemic polio-myelitis with an acute course and a picture similar to Landry's paralysis. Spiller in a rapidly fatal case found destructive changes in the peripheral nerves and alterations in the cell bodies of the ventral horns. He suggests that the toxic agent acts on the lower motor neurones as a whole, and that possibly the reason why no lesions were found in some cases is that the more delicate histological methods were not used. It has much similarity to acute polyneuritis. The view that it is a functional disorder is supported by the study of cases in which no lesion has been found.

**Symptoms.**—Weakness of the legs, gradually progressing, often with tolerable rapidity, is the first symptom. In some cases within a few hours the paralysis of the legs becomes complete. The muscles of the trunk are next affected, and within a few days, or even less in more acute cases, the arms are also involved. The neck muscles are next attacked, and finally the muscles of respiration, deglutition, and articulation. The reflexes are lost, but the muscles neither waste nor show electrical changes. The sensory symptoms are variable; in some cases tingling, numbness, and hyperaesthesia have been present. In the more characteristic cases sensation is intact and the sphincters are uninvolved. The spinal fluid shows no specific features. Enlargement of the spleen has been noted. Bulbar symptoms may be early and there are cases in which the picture has been acute *descending* paralysis. The course of the disease is variable. It may prove fatal in less than two days. Other cases persist for a week or for two weeks. In a large proportion of the cases the disease is fatal, usually by respiratory failure. One patient was kept alive for 41 days by artificial respiration (C. L. Greene).

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**Diagnosis.**—This is difficult, particularly from certain forms of multiple neuritis, and if we include in Landry's paralysis the cases in which sensation is involved distinction between the two affections is impossible. We apparently have to recognize the existence of a rapidly advancing motor paralysis without involvement of the sphincters, without wasting or electrical changes in the muscles, without trophic lesions, and without fever—features sufficient to distinguish it from an acute myelitis or spreading polio-myelitis. It is doubtful, however, whether these characters always suffice to enable us to differentiate the cases of multiple neuritis. The cases of polio-myelitis with the picture of an acute ascending paralysis should not be difficult to recognize during an epidemic.

**Treatment.**—The comfort of the patient should be studied; frequent change of position is useful. An effort should be made to give sufficient food. The bowels should be kept open and retention of urine avoided. Lumbar puncture should be done at once and repeated daily for some days. Sedatives may be required for discomfort or pain. Atropine is advised, especially in view of the danger of excessive bronchial secretion; gr. 1/100 (0.0006 gm.) can be given four times a day. Artificial respiration should be done if necessary, in the hope that the paralysis has reached the height and will soon subside. Convalescence requires no special treatment; massage is useful.

### III. DEGENERATIVE MYELITIS

#### I. COMBINED POSTERO-LATERAL SCLEROSIS

Ataxic Paraplegia (Gowers) : Subacute Ataxic Paraplegia (Russell, Batten and Collier) ; Primary Combined Sclerosis (J. J. Putnam) ; Toxic Combined Sclerosis ; Subacute Combined Degeneration.

**Definition.**—A disorder with symptoms referable to degeneration of the posterior and lateral columns of the cord, occasionally occurring without obvious cause, but most commonly an associated lesion of the cachexias, anæmias, chronic toxæmias and prolonged sub-infections.

**Etiology.**—The disease is most frequent between fifty and sixty, rarely occurs until after thirty, may occur in the aged and affects the sexes equally. The exact etiology is in doubt but some toxic agent is supposed to be responsible. It occurs with pernicious anæmia and sometimes with secondary anæmia, with toxic states as in poisoning from ergot and lead, and in cachectic conditions.

**Pathology.**—The essential change involves the white matter of the cord and brain stem. No increase in neuroglia occurs. The posterior and lateral columns are specially involved and the clinical picture varies with the relative changes in them. There may be areas of degeneration elsewhere, which may take the form of an annular sclerosis, especially in the mid-dorsal region. The nerve cells are affected as a secondary result. Anæmia is frequent and of varying severity and character, being often of the pernicious form. Perhaps the same cause is responsible for the anæmia (when present) and the cord changes.

**Symptoms.**—The onset is insidious and usually with *sensory* disturbances in the fingers and toes; numbness and tingling are common with a variety of other sensations. Loss of pain sense may antedate loss of touch. The sensory disturbance advances from the legs to the trunk and in late stages may be absolute. Loss of muscle sense, vibration sense and of sense of position may be early. Some patients have pains, much like those of tabes, and a girdle sensation is common. *Motor* disturbance may appear early but usually follows the sensory symptoms. The legs are easily tired, the feet drag, and gradually rigidity of the legs comes on. The conditions found will depend on the relative involvement of the posterior and lateral columns. There may be ataxia with the rigidity. The reflexes are exaggerated with an extensor plantar response. As the posterior columns become involved, the spasticity lessens and flaccid paralysis replaces it. The paraplegia may become complete with absence of the knee-jerks, an extensor plantar response, and muscular atrophy. The arms as a rule are not markedly involved and may show sensory changes only. The sphincters are affected in the later stages.

The deep *reflexes* are usually increased in the early and absent in the late stages. The extensor response to plantar stimulation may appear early and is persistent. The skin reflexes are usually increased. Trophic changes in the skin and nails are common. In the late stages mental disturbances may appear. As *anæmia* is common the patients show the usual blood features. The anæmia may follow the cord disease. There is absence of free hydrochloric acid in the stomach. The *course* is variable with an average length of about two years. There are acute cases of short duration and mild forms which persist for years. Remissions occur but the ultimate outcome is fatal.

In the *secondary* variety there may be few or no symptoms in patients long bed-ridden. When fully developed there are (1) muscular hypotony, (2) loss of the knee-jerks, and (3) ataxia, due to involvement of the posterior columns; or (1) muscular hypertony, (2) exaggerated deep reflexes and positive Babinski sign, and (3) motor weakness due to degeneration of the pyramidal tracts (L. F. Barker).

**Diagnosis.**—In the early stages the sensory disturbances should excite suspicion. Syphilis and multiple sclerosis should be excluded. Spinal tumor and tabes may be suggested but a careful study should prevent difficulty. The flaccid form may resemble polyneuritis but the extensor plantar response and absence of tenderness should distinguish them. Friedreich's ataxia should not cause confusion.

**Treatment.**—Any focus of infection should be treated. Iron and arsenic should be given as indicated for anæmia. Special attention should be given to the care of the skin to prevent infection or bed-sores, and infection of the bladder avoided as long as possible. Sedatives may be required for pain. Sleep is sometimes disturbed by spasms for which barbital or pheno-barbital may be given.

**"Central Neuritis."**—This name has been given by Scott to a disease in adults occurring in Jamaica, which perhaps belongs here. The early features are inflammation of the eyes and later changes in the mouth followed by diarrhoea or marked changes in the nervous system. In the latter the first symptoms are sensory disturbances in the feet and legs, followed by inco-ordination and loss of control over the legs. The knee-jerks are absent.

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Death usually occurs from inanition with diarrhoea and true paralysis does not occur. In those who recover there is disturbance of vision, deafness and a peculiar steppage gait. Histologically the nervous system showed general changes, perivascular infiltration, degeneration and fibrosis. The disease suggests some form of toxæmia. The term "central neuritis" was given by Adolf Meyer to a "parenchymatous systemic degeneration, mainly in the nervous system," found in alcoholic, senile and cachectic states and in depressive psychoses at the time of involution. The features are fever, diarrhoea, emaciation, twitching and rigidity of the extremities, and changed reflexes. Mentally there is an anxious agitation with delirium or stupor.

### II. SENILE SPASTIC PARALYSIS

Unlike the Deacon's "Wonderful One-Hoss Shay," the wear and tear incident to daily use tells more on one part of the machine than another. Like Dean Swift "Some go at the top first, others in their legs, others again in both simultaneously." While the whole nervous system may show decay—"the golden bowl broken and the silver cord loosened"—an early sign of old age is the lessening of the control over the muscles, evidenced by tremor and inability to perform the finer movements with the same precision. The gait becomes tottering, the steps uncertain, and at last the use of the legs is lost for purposes of walking, though every muscle group may be put in action. Or one may watch the gradual onset of a spastic paraplegia—a progressive weakness of legs with spasticity and greatly increased reflexes. The steps are short, the feet not lifted from the ground, and the gait uncertain; yet in many cases the strength of the muscles is maintained, and the patient may "keep on his legs" for years. The sphincters are not, as a rule, affected. Arterio-sclerosis is usually present and in premature senility the vessels of the legs may be very stiff and the dorsal arteries of the feet obliterated. Typical intermittent claudication may precede the paraplegia.

### IV. COMPRESSION OF THE SPINAL CORD

(*Compression Myelitis*)

**Definition.**—Interruption of the functions of the cord by slow compression.

**Etiology.**—Caries of the spine, new growths, aneurism, and parasites are the important causes of slow compression. Caries of the spine is in a majority of instances tuberculous and associated with angular curvature. The involvement of the cord is due to pachymeningitis externa, to abscess or in rare cases to direct spicules of bone. There may be a tuberculous pachymeningitis without caries. The injury to the cord may be from interference with the blood supply and œdema, the effects of which are temporary if the condition is relieved. If there is thrombosis of the vessels with resulting necrosis, the damage is permanent. In a few cases it is due to syphilis and occasionally to extension of disease from the pharynx. It is most common in early life, but may occur after middle age. It may follow trauma. Compression may result from aneurism of the thoracic aorta or the abdominal

aorta, in the neighborhood of the celiac axis. Malignant growths of the spine frequently cause a compression paraplegia. A retroperitoneal sarcoma or the growths of Hodgkin's disease may invade the vertebræ. More commonly the involvement is secondary to cancer of the breast. Of parasites, the echinococcus and the cysticercus may occur in the spinal canal.

**Symptoms.**—These may be due to changes in the bones, nerves, and cord. Many symptoms are due to compression of the blood-vessels.

**VERTEBRAL.**—In malignant diseases and in aneurism erosion of the bodies may take place without producing deformity of the spine. Fatal hæmorrhage may follow erosion of the vertebral artery. In caries, on the other hand, it is the rule to find more or less deformity, amounting often to angular curvature. The compression of the cord is rarely if ever the direct result of this bony kyphosis but is due to thickening of the dura, the presence of caseous and inflammatory products between this membrane and the bodies of the vertebræ. The spinous processes of the affected vertebræ are tender on pressure, and pain follows jarring movements or twisting of the spine. There may be extensive tuberculous disease without much deformity, particularly in the cervical region. In the case of aneurism or tumor pain is a constant and agonizing feature.

**NERVE-ROOT SYMPTOMS.**—These result from compression of the nerve roots as they pass out between the vertebræ. In caries, even when the disease is extensive and the deformity great, radiating pains from compression involvement of the roots are rare. Pains are more common in cancer of the spine and may be agonizing. There may be acutely painful areas—the *anæsthesia dolorosa*—in regions of the skin which are anæsthetic to tactile and painful impressions. The nerve trunks are not tender. Trophic disturbances may occur, particularly herpes. Pressure on the ventral roots may give rise to wasting of the muscles supplied by the affected nerves. This is most noticeable in disease of the cervical or lumbar regions.

**CORD SYMPTOMS.**—(a) *Cervical Region.*—The caries may be between the axis and the atlas or between the latter and the occipital bone. In such instances a retropharyngeal abscess may be present, giving rise to difficulty in swallowing. There may be spasm of the cervical muscles, the head may be fixed, and movements may be impossible or cause great pain. In a case in the Montreal General Hospital movement was liable to be followed by transient, instantaneous paralysis of all four extremities, owing to compression of the cord. In one of these attacks the patient died.

In the lower cervical region there may be signs of interference with the cilio-spinal centre and dilatation of the pupils. Occasionally there is flushing of the face and ear of one side or unilateral sweating. Deformity is not so common, but healing may take place with the production of a callus of enormous breadth, with complete rigidity of the neck.

(b) *Thoracic Region.*—The deformity is here more marked and pressure symptoms are more common. The time of onset of the paralysis varies very much. It may be an early symptom, even before the curvature is manifest, and it is noteworthy that Pott first described the disease that bears his name as "a palsy of the lower limbs which is frequently found to accompany a curvature of the spine." More commonly the paralysis is late, occurring many months after the curvature. The paraplegia is slow in its

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development; the patient at first feels weak in the legs or has disturbance of sensation, numbness, tingling, pins and needles. The girdle sensation may be marked, or severe pains in the course of the intercostal nerves. The legs are frequently drawn up, sometimes in spasm, the reflex spinal automatism. Motion is, as a rule, more quickly lost than sensation. The paraplegia is usually of the spastic type, with exaggeration of the reflexes. There is an extensor plantar response. Bastian's symptom—abolition of the reflexes—is rarely met with in compression from caries as the transverse nature of the lesion is rarely complete. The paraplegia may persist for months, or even more than a year, and recovery still be possible.

(c) *Lumbar Region*.—In the lower dorsal and lumbar regions the symptoms are practically the same, but the sphincter centres are involved and the reflexes are not exaggerated.

(d) *Old Lesions of Cord*.—Following trauma in Pott's disease the dura may be much thickened, the cord narrowed and embedded in cicatricial tissue.

*Rapid compression* of the cord usually results from fracture or dislocation. The extent of injury to the cord is variable, depending on the amount of bone injury. Marked compression causes a flaccid paralysis, loss of sensation and reflexes below the lesion and retention of urine followed by incontinence. Bladder infection and bed-sores are common. The outlook is always serious. If there is complete division of the cord, operation should not be done; if there is some function remaining, laminectomy should be done. Operation is indicated if the signs show injury of the cauda equina.

**Diagnosis**.—The X-ray picture is of first importance. The injection of lipiodol into the cisterna magna and by lumbar puncture and finding by the X-rays the distance to which it spreads is useful in determining the site. Caries is by far the most frequent cause of slow compression of the cord, and when there are external signs the recognition is easy. There are cases in which the exudation in the spinal canal between the dura and the bone leads to compression before there are any signs of caries, and if the root symptoms are absent it may be extremely difficult to arrive at a diagnosis. Persistent lumbago is a symptom of importance in masked Pott's disease, particularly after injury. Brown-Séquard's paralysis is more common in tumor and in injuries than in caries. Pressure on the nerve roots, too, is less frequent in caries than in malignant disease. The cervical form of pachymeningitis also produces a pressure paralysis. Following removal of the breast for carcinoma, even after long intervals, recurrence in the vertebræ may cause pressure on the spinal nerves or on the cord. There may be no local recurrence. Neuralgic pains in the neck or back, or in the course of the sciatic, often associated with obscure nervous symptoms, suggesting hysteria, may be present for months before any signs of paralysis or of recurrence elsewhere. The persistence of the pains and their intensity should always arouse suspicion. Finally paraplegia may come on, not often with deformity, and the pains may be of terrible intensity, well deserving the name *paraplegia dolorosa*.

**Treatment**.—In compression by aneurism or metastatic tumors the condition is hopeless. In the former the pains are often not very severe, but in the later morphia is always necessary. Compression by caries is often successfully relieved even after the paralysis has persisted for a long period. When caries is recognized early, rest and support to the spine by various

methods may do much to prevent the onset of paraplegia. When paralysis has occurred, rest with extension gives the best hope of recovery. It is to be remembered that restoration may occur after compression of the cord has lasted for months, or even more than a year. Cases have been cured by recumbency alone, enforced for weeks or months; the extradural and inflammatory products are absorbed and the caries heals. In earlier days brilliant results were obtained in these cases by suspension, a method introduced by J. K. Mitchell in 1826, and pursued with remarkable success by his son, Weir Mitchell. The suspension methods have been superseded by those of hyperextension during recumbency with the application of plaster jackets to hold the body and spine immovable. Forcible correction of the deformity under anæsthesia is not to be recommended. In protracted cases, after these methods have been given a fair trial, operation is usually advisable, and has in many instances been successful. The occurrence of abscess or a sudden increase in deformity with signs of greater pressure indicate the need of operation. In meningeal tumors a laminectomy should be done. In old traumatic lesions operation may be indicated for severe nerve-root pains. The general treatment of caries is that of tuberculosis.

## G. DIFFUSE SCLEROSSES

**General Remarks.**—The supporting tissue of the central nervous system is the neuroglia, derived from the ectoderm. The meninges are composed of true connective tissue derived from the mesoderm, a little of which enters the brain and cord with the blood-vessels. The neuroglia plays the chief part in pathological processes within the central nervous system, but changes in the connective tissue elements may also be important. A convenient division of the cerebro-spinal sclerosis is into degenerative, inflammatory, and developmental forms.

The *degenerative sclerosis* comprise the largest and most important subdivision, in which provisionally the following groups may be made: (a) The common secondary Wallerian degeneration which follows when nerve fibres are cut off from their trophic centres; (b) toxic forms, among which may be placed the sclerosis from lead and ergot, and, most important of all, the sclerosis of the dorsal columns, due in a large proportion of cases to syphilis; (c) the sclerosis associated with change in the smaller arteries and capillaries, met with as a senile process in the convolutions.

The *inflammatory sclerosis* embrace a less important and less extensive group, comprising secondary forms which follow irritative inflammation about tumors, foreign bodies, hæmorrhages, and abscess. Possibly a similar change may follow the primary, acute encephalitis, which Strümpell holds is the initial lesion in the cortical sclerosis so commonly found post mortem in infantile hemiplegia.

The *developmental sclerosis* are believed to be of a purely neuroglial character, and embrace the new growth about the central canal in syringomyelia and, according to French writers, the sclerosis of the dorsal columns in Friedreich's ataxia.

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### MULTIPLE (INSULAR: DISSEMINATED) SCLEROSIS

**DEFINITION.**—A chronic affection of the brain and cord, characterized by localized areas in which the nerve elements are more or less replaced by neuroglia. This may occur in the brain or cord alone, more commonly in both.

**ETIOLOGY.**—It is most common in young persons and in females. The onset is most often between 15 and 30 years of age. Several members in a family may be attacked. It is much less common in the United States than in Great Britain; only 91 cases among 12,000 patients (Collins) against 159 among 2568 cases in three years at the National Hospital, London. The etiology is obscure; trauma, fatigue, cold, exposure, intoxications and infections have all been mentioned. Syphilis plays no part. Many consider some form of infection as responsible and spirochaetes have been described.

**MORBID ANATOMY.**—The sclerotic areas are widely distributed through the white and gray matter. The patches are most abundant in the neighborhood of the ventricles, and in the pons, cerebellum, basal ganglia and the medulla. The cord may be only slightly involved or there may be very many areas throughout its length. The cervical region is apt to be most affected. The nerve roots and the branches of the cauda equina are often attacked. In recently invaded areas there are inflammatory areas showing infiltration with mononuclear cells around the blood-vessels with oedema. Degeneration of the medullary sheaths occurs, with the persistence for some time of the axis-cylinders. There is marked proliferation of the neuroglia, the fibres of which are denser and firmer. Secondary degeneration, although relatively slight, does occur.

**SYMPTOMS.**—When the irregular distribution and number of the lesions are considered, it is easy to realize that the clinical manifestations may show a great variety in character and combination. The early inflammatory and vascular lesions may cause temporary disturbance only, so that the symptoms are of short duration. The tendency is for the sclerosis to increase so that the injury becomes permanent and, therefore, the clinical features become more constant. Two main forms are described; the first (the more frequent) shows acute exacerbations at intervals with alternating quiescent periods, and the second a progressive course. The onset varies; it may be slow and gradual or with some sudden condition which may be temporary. Among the early manifestations are some disturbance of sensation, dragging of one foot, nystagmus, some disturbance of vision, such as diplopia, or of the bladder function (frequency, precipitate micturition, retention, or incontinence). There may be weakness of one or both legs, especially when fatigued, with irregular pains and stiffness. There may be transient dimness of vision or decrease of acuteness of vision, a central scotoma, or retrobulbar neuritis.

As the disease progresses certain features are usually present: (a) *Motor signs.* Weakness in the legs and stiffness, usually increased to a spastic paraplegia with the signs of a pyramidal lesion, increase of the deep reflexes, loss of the abdominal and cremasteric reflexes and an extensor plantar response. The gait may be spastic or also ataxic. The arms are usually affected less than the legs.

(b) *Volitional or intention tremor.* There is no weakness of the arms,

but on attempting to pick up an object there is a trembling or rapid oscillation. A patient may be unable to lift even a glass of water to the mouth. The tremor may be marked in the legs, and in the head, which shakes as he walks. When the patient is recumbent the muscles may be perfectly quiet. On attempting to raise the head from the pillow, trembling at once comes on. (c) *Scanning speech*. The words are pronounced slowly and separately, or the individual syllables may be accentuated. (d) *Nystagmus*, rapid and horizontal, is more common in multiple sclerosis than any other affection of the nervous system. (e) *Ocular signs*. Some of the features have been mentioned. The occurrence of double vision is important. Optic atrophy is common. Pallor of the temporal half of the disk is often seen. (f) *Mental changes*. These are common in the later stages, shown by defective memory, emotional disturbance, and lack of control. Some of these, with vague complaints for which no basis is found, are often responsible for the common error of regarding this disease as hysteria.

*Sensation* is not affected markedly in the majority of cases. The sphincters, as a rule, are unaffected until the later stages. Vertigo is common, and there may be sudden apoplectiform attacks, such as occur in general paresis. The presence of the extensor plantar reflex (Babinski's sign) and absence of the abdominal reflexes are common.

The *course* is variable and the remarkable remissions must be emphasized. Some are steadily progressive. In the later stages the patient usually becomes helpless from the paralysis of the legs, which are often in spastic flexion. Sphincter control is lost and bed-sores are frequent.

**DIAGNOSIS.**—For the early diagnosis three important signs are loss of abdominal reflexes, weakness of the abdominal muscles and pallor of the temporal sides of the optic disks (L. F. Barker). The history of transient disturbances is important, especially diplopia, numbness and weakness. Volitional tremor, scanning speech, and nystagmus form a characteristic symptom-group, but this classical triad is less common than the irregular forms which easily escape recognition. Paralysis agitans, neuro-syphilis, compression of the cord, certain cases of general paresis, and hysteria may simulate the disease very closely. Of all organic diseases of the nervous system disseminated sclerosis in its early stages, is that which is most commonly taken for hysteria (Buzzard, Sr.). The points to be relied upon in the differentiation are spastic weakness of the legs, pallor of the optic disk, absence of the abdominal reflexes, the Babinski sign, nystagmus, bladder disturbances, and the volitional tremor. The tremor in hysteria is not volitional but the diseases may coexist. If in doubt, suspend judgment. Unilateral cases are recorded. The spinal fluid may show the colloidal gold curve with a negative Wassermann reaction.

*Pseudo-sclerosis*—the Westphall-Strümpell disease—is a rare condition simulating multiple sclerosis and not often distinguished from it during life. Mental changes are more pronounced, the tremor is more exaggerated, the nystagmus not always present, and the gait more ataxic. It sets in earlier, sometimes in the first decade, and in a majority of the cases no lesions have been found post mortem.

The **PROGNOSIS** is unfavorable. Ultimately, the patient, if not carried off by some intercurrent affection, becomes bedridden. In 200 cases the average duration was twelve years; 3 recovered (Bramwell).

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**TREATMENT.**—No known treatment has any influence on the progress of sclerosis of the brain. Neither the iodides nor mercury have the slightest effect, but a prolonged course of arsenic may be tried. Energetic treatment with arsphenamine is advised by some. Avoidance of fatigue, physical and mental, is important. In acute stages there should be absolute rest but otherwise activity, short of fatigue, is advisable. Massage, hot baths and muscular re-education help some patients. Benefit has resulted from opening the spinal canal (Elsberg).

**Miliary sclerosis** is a term which has been applied to several different conditions. Gowers mentions a case in which there were grayish red spots at the junction of the white and gray matters, and in which the neuroglia was increased. There is also a condition in which, on the surface of the convolutions, there are small nodular projections, varying from a half to five or more millimetres in diameter.

**Diffuse sclerosis** may involve an entire hemisphere, or a single lobe, in which case the term *sclérose lobaire* has been applied to it by the French. It is not important in general practice, but occurs most frequently in idiots and imbeciles. In extensive cortical sclerosis of one hemisphere the ventricle is usually dilated. The symptoms of this condition depend upon the region affected. There may be a considerable extent of sclerosis without symptoms or much mental impairment. In a majority of cases there is hemiplegia or diplegia with imbecility or idiocy.

**Tuberose Sclerosis.**—Described by Bourneville in feeble-minded children, and regarded as a pathological curiosity, the researches of Vogt, Wolback, Fowler and Dickson and others have shown it to be a definite type of disease, which may sometimes be recognized clinically. Imbecility and epilepsy are present, without, as a rule, paralysis. Anatomically there are remarkable tuberous tumors, embedded in the cortex cerebri, ranging in size from a pea to a walnut, white in color, and of a stony hardness. There is an overgrowth of the neuroglia and of large ganglionic cells. A remarkable peculiarity, which sometimes enables the disease to be recognized, is the occurrence of congenital tumors in other organs, heart, kidneys and skin. Adenoma sebaceum of the face, small, closely-set growth about the nose and cheeks, often with a vascular matrix, is the most common. Renal tumors were found in 19 of 29 cases.

## H. DIFFUSE AND FOCAL DISEASES OF THE SPINAL CORD

### I. TOPICAL DIAGNOSIS

From the symptoms presented by a spinal cord lesion it is possible to determine more or less accurately not only the level but also the transverse extent of the segmental involvement. The effects of injury or disease may be circumscribed and involve the gray matter of the segment or the tracts running through it; it may be more extensive and involve the cord in a given level in its entire transverse extent; finally, there are cases in which only one

lateral half of the cord is implicated. It is well to have a definite routine to follow in making the examinations, for each factor may be helpful in determining the site and character of the lesion. Some of the more important points are the following: (1) *subjective sensations*, particularly the character and seat of pain, if any be present, such as the radiating pains of dorsal root compression; (2) the patient's *attitude*, as the position of the arms in cervical lesions, the character of the respiration, whether diaphragmatic, etc.; (3) *motor symptoms*, the groups of paralyzed muscles and their electrical reaction; (4) the *sensory symptoms*, including tests for tactual, thermic, and painful impressions, for muscle sense, bone sensation, vibration sense, etc.; (5) the condition of the *reflexes*, both the tendon and the skin reflexes as well as those of the pupil, the bladder and rectum, etc.; (6) the surface temperature and condition of the skin, which gives an indication of *vaso-motor* disturbance. The table on pages 921-923 and the figures on pages 928 and 929 will be useful while making an examination.

**Focal Lesions.**—A lesion involving a definite part of the gray matter destroying the cell bodies of the lower motor neurones and leading to degeneration of their axis-cylinder processes, is accompanied by a loss of power to perform certain definite movements. Thus in anterior polio-myelitis the only symptom may be a flaccid paralysis, and the seat of the lesion is revealed by the muscles involved. If from injury or disease a lesion involves more than the gray matter and, for example, if the neighboring fibres of the pyramidal tract be affected there may be in addition a spastic paralysis of the muscles whose centres lie in the lower levels of the cord. The degree of such a paralysis depends upon the intensity of the lesion of the pyramidal tract and may vary from a slight weakness in dorsal flexion of the ankle to an absolute paralysis of all the muscles below the lesion. Again, if the afferent tracts are affected sensory symptoms may be added to the motor palsy. There may be disturbances of pain and temperature sense alone or touch also may be affected. This, however, is rare except in serious lesions. The upper border of disturbed sensation often indicates most clearly the level of the disease, especially when this is in the thoracic region where the corresponding level of motor paralysis is not easily demonstrated. It is unusual for cutaneous anesthesia in organic lesions of the cord to extend above the level of the second rib and the tip of the shoulder, for this represents the lower border of the skin-field of the fourth cervical (see sensory charts), and as the chief centre for the diaphragm lies in this segment, a lesion at this level sufficiently serious to cause sensory disturbances would probably occasion motor paralyses as well and affect respiration. The demonstrable upper border of the anæsthetic field may not quite reach that which represents the level of the lesion. This is due to the functional overlapping of the segmental skin-fields (Sherrington) and applies more to touch than to pain and temperature. There is often a narrow zone of hyperæsthesia above the anæsthetic region.

**Complete Transverse Lesions.**—When the transverse lesion is total and the lower part of the cord is cut off entirely from above, there is complete sensory and motor paralysis to the segmental level of the injury. Certain features are as follows: (1) Total flaccid paralysis of muscles below the level of the lesion. (Spastic paralysis indicates that the lesion is incomplete.) (2) A rapid wasting of the paralyzed muscles with a loss of the faradic excit-

## DISEASES OF THE NERVOUS SYSTEM

ability. (3) The sphincters lose their tone and there is dribbling of urine. (4) There is total anæsthesia to the level of the lesion (the zone of hyperæsthesia is rarer). Riddoch, as a result of the study of cases of complete division of the spinal cord from war injuries, has shown that under favorable conditions the portion of the cord caudal to the lesion not only recovers its reflex functions but becomes highly excitable. Three stages may be recognized: (1) A stage of muscular flaccidity, (2) a stage of reflex activity, and (3) a stage of gradual failure of the reflex function of the isolated portion of the cord.

**Unilateral Lesions** (*Brown-Séquard Paralysis*).—The motor symptoms, which follow lesions limited to one lateral half of the cross section of the spinal cord, are confined to one side of the body; they are on the same side as the lesion. At the level of the lesion, owing to destruction of cell bodies of the lower system of neurones, there will be found flaccid paralysis and atrophy of those muscles whose centres of innervation happen to lie at this level. Owing to degeneration of the pyramidal tract, the muscles whose centres are at lower levels, are also paralyzed, but they retain their normal electrical reactions, become spastic, and do not atrophy to any great degree.

The sensory symptoms are peculiar. On the side of the lesion corresponding to the segment or segments of the cord involved there is a zone of anæsthesia to all forms of sensation. Below this there is no loss in the perception of pain, temperature, or touch. Indeed, hyperæsthesia has been described. Muscle sense is disturbed, and the ability to appreciate the size, consistency, weight, and shape of an object. On the side opposite to the lesion and nearly up to its level there is complete loss of perception for pain and temperature and there may be some dulling of tactile sense as well.

The following table, slightly modified from Gowers, illustrates the distribution of these symptoms in a complete semi-lesion of the cord:

*Cord*

Zone of cutaneous hyperæsthesia. Zone of cutaneous anæsthesia. Lower segment type of paralysis with atrophy.	Lesion.	
Upper segment type of paralysis. Hyperæsthesia of skin. Muscular sense and allied sensations impaired. Reflex action first lessened and then increased. Surface temperature raised.		Muscular power normal. Loss of sensibility of skin to pain and temperature. Muscular sense normal. Reflex action normal. Temperature same as that of above lesion.

It is common in syphilitic diseases of the cord, tumors and stab-wounds, and is not infrequently associated with syringomyelia and hemorrhages into the cord. It is only in exceptional cases, of course, that the lesion is absolutely limited to the hemi-section of the cord and the symptoms consequently may vary somewhat in degree.

**Lesions of the Conus medullaris and Cauda equina.**—The chief lesions of this region are (1) fractures and dislocations, (2) myelitis, (3) tumors, (4) gunshot wounds, and (5) neuritis of the nerves of the cauda.

(1) **CONUS ALONE.**—It may be in the seat of a tumor or a focal myelitis or hæmorrhage, and it has been damaged in a lumbar puncture. The features are characteristic—paralysis of the rectum and bladder, with the “riding-breeches anæsthesia” of the perineum, scrotum, penis, and postero-internal aspects of the thigh and absence of the ankle jerk. There is less pain than in caudal lesions: the disturbance of sensation is bilateral.

(2) The **EPICONUS** may be involved alone, leading to degenerative atrophy of the muscles innervated by the sacral plexus, particularly the peronei and the glutei. “If the lesion be limited to the grey matter of the epiconus, the Achilles reflex is abolished, but the knee-jerk can be elicited and the sphincters remain unaffected” (Barker).

(3) **CAUDA EQUINA.**—An unusual number of cases followed bullet and shell wounds in the late war. The picture varies with the level of the lesion, from complete paralysis of all the muscles of the legs with anæsthesia, including the genitals, but if below the second sacral roots, there is no paralysis of the lower limbs, but there is the typical saddle-shaped anæsthesia. The caudal lesions are more often unilateral, and the neuralgic pains are more severe.

Of tumors of the cauda mention must be made of the diffuse giant tumors described by Collins and Elsberg, with well marked caudal and conus symptoms. There is also a remarkable *neuritis* in which the caudal roots are swollen and the nerves degenerated, in association with a high grade of local arterio-sclerosis. The symptoms in the cases reported by Kennedy and Elsberg were pain, sphincter involvement, and sensory changes in the sacral roots.

## II. AFFECTIONS OF THE BLOOD-VESSELS

### I. CONGESTION

Apart from actual myelitis, we rarely see congestion of the spinal cord, and, when we do, it is usually limited either to the gray matter or to a definite portion of the organ. The white matter is rarely found congested, even when inflamed. The gray matter often has a reddish pink tint, but rarely a deep reddish hue, except when myelitis is present. If we know little anatomically of congestion of the cord, we know less clinically, for there are no features in any way characteristic of it.

### II. ANÆMIA

So, too, with this state. There may be extreme grades of anæmia without symptoms. There is no reason to suppose that such sensations as heaviness in the limbs and tingling are especially associated with anæmia.

Profound anæmia follows ligation of the aorta. Within a few moments after the application of the ligature paraplegia came on (Herter). Paralysis of the sphincters occurred, but less rapidly. Observations made by Halsted on occlusion of the abdominal aorta in dogs showed that paraplegia occurs

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In a large percentage of cases, many of which, however, may recover as the collateral circulation is established. In the fatal cases Gilman found extensive alterations in the cell bodies of the lower part of the cord with degenerations. This is of interest in connection with the occasional rapid development of a paraplegia after profuse hæmorrhage, usually from the stomach or uterus. It may come on at once or at the end of a week or ten days, and is probably due to an anatomical change in the nerve elements similar to that produced in Herter's experiments. The degeneration of the cord in pernicious anæmia has been described.

### III. EMBOLISM AND THROMBOSIS

Blocking of the spinal arteries by emboli rarely occurs. Thrombosis of the smaller vessels in connection with endarteritis plays an important part in many of the acute and chronic changes in the cord.

### IV. ENDARTERITIS

It is remarkable how frequently in persons over fifty the arteries of the spinal cord are found sclerotic. The following forms may be met with: (1) A nodular peri-arteritis or endarteritis associated with syphilis and sometimes with gummata of the meninges; (2) an arteritis obliterans, with great thickening of the intima and narrowing of the lumen, involving chiefly the medium and larger-sized arteries. Miliary aneurisms or aneurisms of the larger vessels are rarely found in the spinal cord. Attacks of transient paraplegia may be due to spasm or other changes in the vessels of the cord. In the remarkable neuritis of the cauda equina described by Kennedy and Elsberg there is marked sclerosis of the arteries.

### V. HÆMORRHAGE INTO THE SPINAL MEMBRANE; HÆMATOCHISIS

In meningeal "apoplexy," as it is called, the blood may lie between the dura mater and the spinal canal—extra-meningeal hæmorrhage—or within the dura mater—intra-meningeal hæmorrhage.

**Extra-meningeal hæmorrhage** occurs usually as a result of traumatism. The exudation may be extensive without compression of the cord. The blood comes from the large plexuses of veins which may surround the dura. The rupture of an aneurism into the spinal canal may produce extensive and rapidly fatal hæmorrhage.

**Intra-meningeal hæmorrhage** is a less frequent result of trauma, but in general is perhaps rather more common. It is rarely extensive from causes acting directly on the spinal meninges themselves. Scattered hæmorrhages are not infrequent in the acute infectious fevers, and there may be much extravasation in malignant small-pox. It may be into the theca alone and along the spinal nerve roots. Bleeding may occur also in death from convulsive disorders, such as epilepsy, tetanus, and strychnia poisoning, and has been recorded with difficult parturition and in purpura. The most extensive hæmorrhages occur from rupture of an aneurism at the base of the brain, either of the basilar or vertebral artery. In ventricular hæmorrhage the blood may pass

from the fourth ventricle into the spinal meninges. In cranial fractures, particularly of the base of the skull, the resultant hæmorrhage almost always finds its way into the subarachnoid space about the cord and may be demonstrated by the withdrawal of bloody fluid by a lumbar puncture. The procedure is of considerable diagnostic value. On the other hand, hæmorrhage into the spinal meninges may possibly ascend into the brain.

**Symptoms.**—The symptoms in moderate grades may be slight and indefinite. The spinal features suggest lumbar puncture and the nature of the fluid, flowing under pressure, determines the presence of hæmorrhage. In the non-traumatic cases the hæmorrhage may either come on suddenly or after a day or two of uneasy sensations along the spine. As a rule, the onset is abrupt, with sharp pain in the back and symptoms of irritation in the course of the nerves. There may be muscular spasms, or paralysis may come on suddenly, either in the legs alone or both in the legs and arms. In some instances the paralysis develops more slowly and is not complete. There are no signs of cerebral disturbance. The clinical picture varies. If the hæmorrhage is in the *lumbar* region, the legs alone are involved, the reflexes may be abolished, and the action of the bladder and rectum is impaired. If in the *thoracic* region, there is more or less complete paraplegia, the reflexes are usually retained, and there are signs of disturbance in the thoracic nerves, such as girdle sensations, pains, and sometimes eruption of herpes. In the *cervical* region the arms as well as the legs may be involved; there may be difficulty in breathing, stiffness of the muscles of the neck, and occasionally pupillary symptoms. In a case of influenza-pneumonia in the recent epidemic there was bilateral spastic rigidity associated with extensive hæmorrhage into the theca spinalis and along the nerve roots. There was no free blood in the canal. Branson reports two cases, probably influenza, with bloody fluid (40-50 c. c.) withdrawn under considerable pressure. The spinal symptoms were slight and both patients recovered.

The *diagnosis* from hæmorrhage into the cord is made by the signs of irritation preceding the paralysis, which is usually less marked. The study of the spinal fluid distinguishes it from meningitis. The *prognosis* depends much upon the cause. Recovery may take place in the traumatic cases and in those associated with the infectious diseases. The *treatment* depends largely on the cause. Lumbar puncture should be repeated as indicated.

## VI. HÆMORRHAGE INTO THE SPINAL CORD: HÆMATOMYELIA

Most frequently a result of traumatism, intraspinal hæmorrhage is naturally more common in males and during the active period of life. In some cases no cause can be found. Cases have followed cold or exposure; it occurs also in tetanus and other convulsive diseases, and hæmorrhage may be associated with tumors, syringomyelia or myelitis. A direct injury to the spine is by far the most common cause. Acute flexure of the neck, often without fracture or dislocation of the vertebræ, is the most common form of accident. There were many such cases during the war. The level of the lesion, for this reason, is most frequently in the lower cervical region.

**Anatomical Condition.**—The extent of the hæmorrhage may vary from a small focal extravasation to one which finds its way in columnar fashion

a considerable distance up and down the cord. The bleeding primarily takes place into the gray matter, and this as a rule suffers most, but the surrounding medullated tracts may be thinned out and lacerated.

**Symptoms.**—As one side of the cord is usually involved more than the other, the Brown-Séquard syndrome is common. The symptoms are sudden in onset, and leave the patient with hyperæsthesia and a paralysis which becomes spastic and is most marked on one side, while anæsthesia, chiefly to pain and temperature, is most marked on the opposite side of the body. Often a most distressing hyperæsthesia, usually a "pins and needles" sensation, may be present for many days, but there is rarely any acute pain of the radiating or root type. As hæmatomyelia is most frequent in the lower cervical region, in addition to the symptoms just mentioned a brachial type of palsy is commonly seen, with flaccid and atrophic paralysis of the muscles innervated from the lowest cervical and first thoracic segments. The hæmorrhage may occur in segments farther down the cord, the lumbar enlargement being affected next in frequency to the lower cervical. The segmental level of the paralysis necessarily would vary accordingly.

The condition may prove rapidly fatal, particularly if the extravasation is bilateral and extends high enough in the cord to involve the centres for the diaphragm. More frequently there is a more or less complete recovery with a residual palsy of the upper extremity and a partial anæsthesia, corresponding to the level of the lesion, and some spasticity of the leg.

**Diagnosis.**—In the traumatic cases this is comparatively easy, and it is important to recognize them, as they are often needlessly subjected to operation under the belief that they are instances of acute compression. Myelitis and polio-myelitis have to be excluded. The residual symptoms in old cases may closely simulate those in syringomyelia.

**Treatment.**—Absolute rest is important and the patient should be disturbed as little as possible. Special care must be given the skin to prevent bed-sores and to the bladder to prevent cystitis. If lumbar puncture is done for diagnosis, the least amount necessary should be removed. Treatment of the paralyzed parts should not be begun for six weeks after the hæmorrhage, when electricity, gentle massage, and passive movements are indicated.

### III. TUMORS OF THE SPINAL CORD AND ITS MEMBRANES

#### 1. SYRINGOMYELIA (GLIOMA, GLIOMATOSIS)

**Definition.**—A gliosis about the central canal, either forming a local tumor, or more often a diffuse growth associated with cavity formations, extending lengthwise, and sometimes communicating with the central canal.

Dilatation of the central canal—hydromyelus—which must be distinguished from syringomyelia, is met with as a congenital anomaly; only in a few instances do the cavity formations of syringomyelia represent the distended canal itself.

**Morbid Anatomy.**—The lower cervical and upper dorsal regions are the

usual seat. There are: (1) either a diffuse gliosis or at one level a definite tumor from which the growth extends for some inches, causing enlargement of the cord. (2) Tube-like cavities, extending for a variable distance, usually in the dorsal aspect and sometimes involving only one cornu. The processes leading to the formation of the cavities are various, such as hæmorrhage and thrombotic degenerations, evidences of which may be present. The wall of the tubes may be smooth and lined with ependymal cells. (3) Degenerative changes in other parts of the cord due to pressure.

**Symptoms.**—Men are more often affected, 133 of 190 cases collected by Schlesinger. The disease begins, as a rule, before the thirtieth year. The symptoms vary with the seat and extent of the disease. A typical case beginning in the lower cervical region presents the following features: (1) Lower motor neurone involvement, with a progressive atrophy of the muscles of the hands and arms, and sometimes fibrillary tremors, so that the Aran-Duchenne disease is suspected. The typical claw-hand may exist. As the disease progresses, there is degeneration of the pyramidal tracts with a spastic paraplegia, so that the picture suggests amyotrophic lateral sclerosis.

(2) *Sensory* changes; (*a*) pains of the nerve-root type, chiefly in the arms; (*b*) the syringomyelic *dissociation of sensation*, in which the sense of touch is retained, while those of heat and of pain are lost. The muscular sense is not disturbed. The loss of temperature sense may be early, and a patient's fingers may be burnt by a cigarette.

(3) *Trophic* changes, as destructive whitlows, with atrophy of the terminal phalanges (Morvan's disease), vaso-motor swelling of the hands, thickening of the skin, sweating, and arthropathies, which latter occur in about 10 per cent. of the cases. While this is the common form, there may be no disturbance of sensation for years, only the amyotrophic type of paralysis; there may be general anæsthesia to pain and temperature, with very little motor disturbance; and there is a form with bilateral spastic diplegia.

Marked scoliosis may be present, a feature not easily explained. The analgesia and loss of thermic sense are due to involvement of the periependymal gray matter and the posterior horns. The tactile sensations travel in the postero-lateral regions of the cord which are rarely involved. Disturbance of the cervical sympathetic is common. With higher involvement there may be nystagmus, diplopia, or atrophy of the tongue.

The *diagnosis* is easy in well pronounced cases, but when the motor features predominate, it may not be possible to distinguish the disease from amyotrophic muscular paralysis. With the widespread anæsthesia hysteria is simulated; while the combination of anæsthesia and loss of the finger tips may suggest leprosy. A cervical rib may give very similar features; the two conditions may coexist. In a few instances the gliosis extends to the medulla with the production of bulbar symptoms. The *course* is variable and there may be no change for long periods. The tendency is to slow progression.

**Treatment.**—Care should be taken to avoid injury and local infection. Active treatment by mercury, given by inunction, and iodide is advised. X-ray exposures over the spine have been followed by improvement. Laminectomy with drainage of the cavity fluid has been of benefit, but there is danger from an anæsthetic. Cervical puncture has been done.

## DISEASES OF THE NERVOUS SYSTEM

### II. TUMORS OF THE CORD AND MENINGES

Tumors may be situated within the membranes (intra-dural) and in the spinal cord (intra-medullary) or outside it (extra-medullary). Extra-dural tumors usually involve the vertebræ and are commonly metastatic carcinomas. The intra-medullary tumors are usually gliomas. A majority are extra-medullary, and originate on the dura or pia in the blood-vessels or on the nerve-roots. Schlesinger's tabulation of 400 cases shows that the growths in order of frequency are tubercle, fibroma and syphiloma. Rarer forms are lipoma, psammoma, neuroma, myxoma and angioma. A few cases of aneurism and echinococcus cyst have been reported. There is an occasional diffuse sarcomatous tumor which tends to spread up and down.

**Symptoms.**—Localized persistent *pain*, perhaps with occasional remission in severity is an important and usually the first symptom. Subjective sensory disturbances follow with many varieties of paræsthesia. The pain, due to root involvement, is on the same side as the lesion, but paræsthesia, due to compression, may be on either or both sides. In some cases with a small tumor from the pia or arachnoid on the anterior aspect or posterior between the nerve roots, pain may be of late occurrence. (1) *Irritation*—sensory and motor. Pressure on the posterior roots causes pain, unilateral or bilateral, at the level of the distribution of the nerves. Hyperæsthesia with a sense of burning is common. In the cervical region the sympathetic fibres may be involved. Only in a few cases are sensory features absent. *Motor* irritation due to pressure on the anterior roots and antero-lateral columns causes spontaneous spasms of the muscles, rarely of the arms, very often of the legs, and they constitute an important sign. Suddenly, without the patient's knowledge, the legs are drawn up, sometimes in pain, the thighs flexed on the abdomen, the legs on the thighs and foot, and especially the big toe on the ankle. It is a reflex of spinal automatism similar to that described by Sherrington in the decerebrated animal. It is the "defensive" reflex of Babinski, but Sherrington's term is preferable. It may be excited by stimulating the skin of the leg or foot, but the important point is the automatic type of the reflex and its significance as a sign of pressure irritation on the cord, at any stage early or late of the process.

(2) *Compression*.—Anæsthesia may occur in the region of distribution of the nerve-root or roots involved; atrophy of the muscles may follow pressure on the anterior roots. Pressure on the cord may produce the features of a hemi-lesion with a Brown-Séquard syndrome. Gradually, after months or even years, the compression is complete with a spastic paraplegia and all the features of a spinal automatism. All stages from nerve-root irritation to a total transverse lesion may be followed through a period of months or years.

The situation of the growth is determined by the root-levels involved, and it is to be remembered that the tendency is usually to locate it below the actual situation. The X-rays are often of great value in determining the nature of the pressure, particularly in excluding disease of the vertebræ.

**Spinal Fluid.**—From an extensive study Sprunt and Walker describe two forms of xanthochromia, in one of which the color is due to dissolved hæmoglobin, the fluid does not coagulate, and the amount of globulin is small. This is more common with brain tumor. In a larger group the fluid is clear yel-

low, coagulates, and has a large amount of globulin and no hæmoglobin—the so-called Froin syndrome; and is a compression sign, associated with the isolation of a cul-de-sac, in which the fluid stagnates. It suggests spinal tumor or intra-dural inflammation. Lumbar puncture may give a clear normal fluid. The leucocytes may be increased; the chief interest is the occurrence of xanthochromia.

**Diagnosis.**—The order of progress is usually pain, paræsthesia, paralysis. Most tumors are lateral. Ventral or ventro-lateral tumors are most likely to give indefinite features. Posterior tumors have root pains and posterior column disturbances particularly. In the segment diagnosis the level of the sensory change is the most important point. When constant and severe root pains are associated with a progressive paralysis, the diagnosis may be easily made. Caries may cause identical symptoms, but the radiating pains are rarely so severe. Cervical meningitis simulates tumor very closely, and in reality produces identical effects, but the slow progress and the bilateral character from the outset may be sufficient to distinguish it. Syphilitic meningo-myelitis may resemble tumor and present radiating pains, a sense of constriction, and progressive paralysis. Syringomyelia may give a similar picture. A radiogram may be of aid in case the vertebræ are infiltrated by the growth. The nature of the tumor can rarely be indicated with precision. With a marked syphilitic history gumma may be suspected, or, with coexisting tuberculous disease, a solitary tubercle. Tumors of the cord itself cause loss of power and sensation below the lesion. There may be paræsthesia before the anesthesia or pains in the legs. Occasionally there is dissociation of sensation which is an aid in the diagnosis of extra-medullary tumors. The picture varies greatly depending on the parts of the cord first and most involved.

**Treatment.**—It is difficult to say which rouses the greater admiration—the brilliant diagnosis of the clinician or the technique of the physiological surgeon, the combination of which enabled Gowers and Horsley to remove, for the first time, and with permanent success, a tumor of the spinal cord. The report of this case should be read to his class by every teacher of neurology (Medico-Chir. Soc. Trans., London, LXXI, 1888). In syphiloma recovery is possible, even after complete paraplegia. The hopeful cases are the isolated growths springing from the membranes, and operation has been followed by an ever increased percentage of recovery.

## I. DIFFUSE AND FOCAL DISEASES OF THE BRAIN

### I. TOPICAL DIAGNOSIS

In many regions disease may exist without causing symptoms—the so-called *silent areas*. Other areas at once give symptoms. These are the cortical motor centres and the associated sensory centres, the speech centres, the centres for the special senses, and the tracts which connect these cortical areas with each other and with other parts of the nervous system. The following is a brief summary of the effects of lesions from the cortex to the spinal cord:

## DISEASES OF THE NERVOUS SYSTEM

**The Cerebral Cortex.**—(a) *Destructive lesions* of the motor cortex cause *paralysis* in the muscles of the opposite side of the body. The paralysis is at first flaccid, later spastic, the extent depending upon that of the lesion. It is apt to be limited to the muscles of the head or of an extremity, giving rise to the cerebral monoplegias. One group of muscles may be more affected than others, especially in lesions of the highly differentiated area for the upper extremity. It is uncommon to find all the muscle groups of an extremity equally involved in cortical monoplegia. In small bilateral symmetrical lesions monoplegia of the tongue may result without paralysis of the face. A lesion may involve centres lying close together or overlapping one another, thus producing associated monoplegias—e. g., paralysis of the face and arm, or of the arm and leg, but not of the face and leg without involvement of the arm. Very rarely the whole motor cortex is involved, causing paralysis of the opposite side—cortical hemiplegia.

Adjoining and posterior to the motor area is the region of the cortex in which the impulses concerned in general bodily sensation (cutaneous sensibility, muscle sense, visceral sensations) first arrive (the somæsthetic area). Combined with the muscular weakness there is usually some disturbance of sensations, particularly of those of the muscular sense. In lesions of the superior parietal lobe the stereognostic sense is very often affected. For example, when a coin or a knife is placed in the hand of the affected limb, the patient's eyes being closed, it is not recognized, owing to inappreciation of the form and consistence of the object, and this even though the slightest tactile stimulus applied to the fingers or surface of the hand is felt and may be correctly localized. The sense of touch, pain, and temperature may be lowered, but not markedly unless the superior and inferior parietal lobules are involved in subcortical lesions. Paræsthesias and vaso-motor disturbances are common accompaniments of paralyzes of cortical origin.

(b) *Irritative lesions* cause localized spasms. The most varied muscle groups corresponding to particular movement forms may be picked out. If the irritation be sudden and severe, typical attacks of Jacksonian epilepsy may occur. These are often preceded and accompanied by subjective sensory impressions. Tingling or pain, or a sense of motion in the part, is often the *signal symptom* (Seguin), and is of aid in determining the seat of the lesion.

When lesions are both destructive and irritative, there are combinations of the symptoms produced by each. For instance, certain muscles may be paralyzed, and those represented near them in the cortex may be the seat of localized convulsions, or the paralyzed limb itself may be at times subject to convulsive spasms, or muscles which have been convulsed may become paralyzed. The close observation of the sequence of the symptoms in such cases often makes it possible to trace the progress of a lesion involving the motor cortex. The most frequent cause is a developing tumor, though sometimes local thickenings of the membranes of the brain, small abscesses, minute hæmorrhages, or fragments of a fractured skull are responsible.

**Centrum semiovale.**—Lesions may involve either projection fibres (motor or sensory) or association fibres. If the involvement of the motor path causes paralysis, this has the distribution of a cortical palsy when the lesion is near the cortex, and of a paralysis due to a lesion of the internal capsule when it

is near that region. Other systems of fibres running in the centrum semiovale may be involved causing sensory disturbances—hemianæsthesia and hemianopia—and if the lesion is in the left hemisphere, one of the different forms of aphasia may accompany the paralysis.

Two other features may be associated with a cortical or indeed with any lesion. *Neighborhood symptoms* are produced by pressure. A tumor may cause disturbance of function in adjacent centres, or interrupt motor or sensory paths. A hæmorrhage often causes transient symptoms which clear up after the clot shrinks. Transient disturbances of the speech centres and temporary involvement of the paths in the internal capsule are common effects. *Distal symptoms* are produced in two ways. The pressure of a tumor in the frontal lobe may influence the function of the motor centres or a pituitary growth may affect far distant parts, with localizing symptoms.

*Shock symptoms* (which were much studied during the war) arise from functional disturbance of parts distant from the site of the lesion. A blow in the head may abolish the knee-jerk; transient aphasia may be caused by a fall on the right side of the head. The loss of consciousness in apoplexy may be due in part to the shock of the stroke. In the psychic side of war this shock action in causing local or widespread loss of function played an important rôle. The deleterious effect on neurones or centres far removed from the site of the injury is called *diaschisis* by von Monakow.

**Corpus striatum.**—Nothing is known of the functions of the caudate nucleus. The progressive lenticular degeneration (Wilson's disease) is described among the familial nervous affections. The globus pallidus, part of the lenticular nucleus, is involved in paralysis agitans and in Huntington's chorea (Ramsay Hunt).

**Corpus callosum.**—It may be absent congenitally. Though often involved in tumors, characteristic symptoms are rare. One of special interest has been noted by Liepmann in connection with apraxia. The left half of the brain is the dominant partner (as more than 90 per cent. of persons are right-handed) in our manual activities, but through the fibres of the corpus callosum it has guiding influences on the movements controlled by the right hemisphere. Thus a lesion of the left cerebrum above the capsule may cause apraxia of the left arm by cutting the callosal fibres through which influences pass from the left to the right arm centres. The anomalous features of right hemiplegia or monoplegia with apraxia of the non-paralyzed arm are suggestive of a callosal lesion.

**The Thalamus.**—Much knowledge of its function has been obtained by a study of local lesions. It is an important sub-station in the sensory path, and, as Nothnagel showed, it is the lower reflex centre for the emotional movements of laughing and crying; and lesions of this part have long been known to be associated with athetoid and choreic movements.

The *thalamic syndrome* consists of: (1) Contra-lateral hemianæsthesia, sometimes with severe pains; (2) irregular movements—ataxic, choreic, or athetoid; and, (3) as the lesion progresses, hemi-paresis, but the plantar reflex may remain flexor. Lesions of the posterior third may involve the optic radiations causing bilateral homonymous hemianopia. Control of the voluntary movements with loss of the mimic associated movements of the lower half of the face in laughing and crying suggests a thalamic lesion.

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**Internal Capsule** (Fig. 18).—Through this pass within a rather narrow area all, or nearly all, of the projection fibres (motor and sensory) which are connected with the cerebral cortex. It is divided into an anterior limb, a knee, and a posterior limb, the latter consisting of a thalamo-lenticular portion (anterior two-thirds) and a retro-lenticular portion (posterior third). The principal bundle passing through the anterior limb of the capsule is that connecting the frontal gyri and the medial bundle in the base of the peduncle (crus) with the nuclei of the pons. These fibres are centrifugal, and innervate chiefly the lower motor nuclei governing bilaterally innervated muscles, especially of the eyes, head, neck, and probably those of the mouth, tongue, and larynx. In lower horizontal planes these fibres are situated near the knee of the capsule. It is the region of the knee of the capsule which transmits especially the fibres passing from the cerebral cortex to the nuclei of the facial, hypoglossal, and third nerves. The path which supplies the nuclei governing the muscles used in speech passes through the knee.

The pyramidal tract goes through the thalamo-lenticular portion of the capsule. The motor fibres are arranged according to definite muscle groups, or rather movement forms, those for the movements of the arm being anterior to those for the leg. The number of fibres for a given muscle group corresponds rather to the degree of complexity of the movements than to the size of the muscles concerned. Thus the areas for the fingers and toes are relatively large.

The fibres to the somæsthetic area of the cortex—that is, those from the centro-lateral group of nuclei of the thalamus and the tegmental radiations—carrying impulses concerned in general bodily sensation, pass upward through the posterior part of the thalamo-lenticular portion of the capsule. Some of these fibres pass through the anterior two-thirds of the posterior limb alongside of the fibres of the pyramidal tract.

Through the retro-lenticular portion of the posterior limb, opposite the posterior third of the lateral surface of the thalamus, pass (1) the fibres carrying impulses concerned in the sensations of the opposite visual field (optic radiation from the lateral geniculate body to the visual sense area in the occipital cortex); (2) the fibres carrying impulses concerned in auditory sensations (radiation from the medial geniculate body to the auditory sense area in the cortex of the temporal lobe); (3) the fibres (probably centrifugal) connecting the cortex of the temporal lobe with the nuclei of the pons.

With this preliminary knowledge concerning the internal capsule, it is not difficult to understand the symptoms which result when it is diseased.

Since here all the fibres of the upper motor segment are gathered together in a compact bundle, a lesion in this region is apt to cause complete hemiplegia of the opposite side, followed later by contractures; and if the lesion involves the hinder portion of the posterior limb there is also hemianæsthesia, including even the special senses. As a rule, however, lesions of the internal capsule do not involve the whole structure. The disease usually affects the anterior or posterior portions, and even in instances in which at first the symptoms point to total involvement there is a disappearance often of a large part of the phenomena after a short time. Thus, when the pyramidal tract is destroyed (lesion of the thalamo-lenticular portion of the capsule) the arm may be affected more than the leg, or *vice versa*. The facial paralysis is

usually slight, though if the lesion be well forward in the capsule the paralysis of the face and tongue may be marked.

The bilaterally innervated muscles of the upper face, of mastication, of deglutition, phonation, and of the trunk muscles are very slightly involved. The patient can wrinkle the forehead, and close the eye on the affected side, but the muscles may be weak, as shown by lessened respiratory movement on the paralyzed side.

Hemianæsthesia alone without involvement of the motor fibres is rare. There is usually also at least partial paralysis of the leg. When the retrolenticular portion of the capsule is destroyed the hemianæsthesia is accompanied by hemianopia, disturbances of hearing, and sometimes of smell and taste. The occurrence of hemianæsthesia with pain, hemichorea, marked tremor, or hemiathetosis—thalamic syndrome—after a capsular hemiplegia points to the involvement of the thalamus or of the hypothalamic region.

Charcot and others have described cases in which as a result of disease of the internal capsule there has been paralysis of the face and leg without involvement of the arm. In such instances the lesion is linear, extending from the posterior part of the anterior limb of the internal capsule backward and lateralward to the leg region in the posterior limb of the capsule, the region for the arm escaping.

Capsular lesions when pure are not usually accompanied by aphasic symptoms, alexia, or agraphia. A "subcortical" motor aphasia may result if the lesion is bilateral, as in pseudo-bulbar paralysis, or if on the left side it is so extensive as to destroy the fibres connecting Broca's convolution with the opposite hemisphere, as well as the pyramidal fibres on the same side.

**Crura (Cerebral Peduncles).**—From this level through the pons, medulla, and cord the upper and lower motor segments are represented, the first by the fibres of the pyramidal tracts and by the fibres which go from the cortex to the nuclei of the cerebral nerves, the latter by the motor nuclei and the nerve fibres arising from them. Lesions often affect both motor segments, and produce paralysis having the characteristics of each. Thus a single lesion may involve the pyramidal tract and cause a spastic paralysis on the opposite side of the body, and also involve the nucleus or the fibres of one of the cerebral nerves, and so produce a lower segment paralysis on the same side as the lesion—crossed paralysis. In the crus the third and fourth cerebral nerves run near the pyramidal tract, and a lesion of this region is apt to involve them or their nuclei, causing partial paralysis of the muscles of the eye on the same side as the lesions, combined with a hemiplegia of the opposite side (Weber-Gubler syndrome) (Fig. 15, 3).

The optic tract also crosses the crus and may be involved, giving hemianopsia in the opposite halves of the visual fields.

If the tegmentum be the seat of a lesion which does not involve the base of the peduncle (or pes) there may be disturbances of cutaneous and muscular sensibility, ataxia, disturbances of hearing, or oculo-motor paralysis. An oculo-motor paralysis of one side, accompanied by a hemiataxia of the opposite side, appears to be especially characteristic of a tegmental lesion (Benedikt's syndrome). Or there may be with the crossed paralysis the features of cerebellar ataxia (Nothnagel's syndrome).

**Corpora quadrigemina.**—Anatomical studies point to the view that the superior colliculus (anterior quadrigeminal body) represents the most important subcortical central organ for the control of the eye-muscle nuclei. This is supported to a certain extent by clinical evidence. Sight is only slightly, if at all, disturbed when the superior colliculus is destroyed. The pupil is usually widened, and the pupillary reaction, both to light and on accommodation, interfered with. Apparently actual paralysis of the eye muscle does not occur unless the nucleus of the third nerve ventral to the aqueduct be also injured.

The inferior colliculus (posterior quadrigeminal body) is an important way-station in the auditory conduction-path. A large part of the lateral lemniscus ends in its nucleus, and from it emerge medullated fibres which pass through the brachium quadrigeminum inferius to the medial geniculate body. Thence a large bundle runs through the retro-lenticular portion of the internal capsule to the auditory sense area in the cortex of the temporal lobe. In 9 of 19 tumors of this region collected by Weinland there were auditory disturbances.

Since the central auditory path of each side receives impulses from both ears, lesion of the colliculus on one side may dull the hearing on both sides, though the opposite ear is usually the more defective. Lesion of the inferior colliculus may be accompanied by disturbance of mastication, owing to paralysis of the descending (mesencephalic) root of the trigeminus. The fourth nerve may also be involved. The ataxia which sometimes accompanies lesions of the corpora quadrigemina is probably to be referred to disturbance in conduction in the medial lemniscus.

**Pons and Medulla oblongata.**—Lesions involving the pyramidal tract, together with any one of the motor cerebral nerves of this region, cause crossed paralysis—*hemiplegia alternans*. A lesion in the lower part of the pons causes a lower-segment paralysis of the face on the same side (destruction of the nucleus of the facial nerve or of its root fibres) and a spastic paralysis of the arm and leg on the opposite side (injury to pyramidal tract) (Fig. 15, 4). This is referred to as the alternate hemiplegia or the Millard-Gubler type. The abducens, the motor part of the trigeminus, and the hypoglossus nerves may also be paralyzed in the same manner. When the central fibres to the nucleus of the hypoglossus are involved a peculiar form of anarthria results. If the nucleus itself be diseased, swallowing is interfered with.

When the sensory fibres of the fifth nerve are interrupted, together with the sensory tract (the medial lemniscus or fillet) for the rest of the body, which has already crossed the middle line, there is a crossed sensory paralysis—i. e., disturbed sensation in the distribution of the fifth on the side of the lesion, and of all the rest of the body on the opposite side—*hemi-anæsthesia cruciata*.

A paralysis of the external rectus muscle of one eye and of the internal rectus of the other eye (conjugate paralysis of the muscles which turn the eyes to one side), in the absence of a "forced position" of the eyeballs, is highly characteristic of certain lesions of the pons. In such cases the internal rectus may still be capable of functioning on convergence, or when the eye to which it belongs is tested independently of that in which the external rectus is paralyzed. This form, known as the Foville type of hemi-

plegia alternans, is found, as a rule, only when the lesion lies just in front of the abducens or involves the nucleus itself, or includes, besides the root fibres of the abducens, that portion of the formatio reticularis that lies between them and the fasciculus longitudinalis medialis (von Monakow). The facial nerve is often involved in these paralyses.

In lesions of the pons the patient often has a tendency to fall toward the side on which the lesion is, probably on account of implication of the middle peduncle of the cerebellum (brachium pontis). Still more frequent is the simple motor hemiataxia consequent upon lesion of the medial lemniscus, and perhaps of longitudinal bundles in the formatio reticularis. This is often accompanied by a dissociated sensory disturbance, pain and temperature being affected, while touch remains normal. The muscular sense may also be involved. Only when the lesion is very extensive are there disturbances of hearing (involvement of the lateral lemniscus or corpus trapezoideum).

So small is the space in which important paths and nuclei are crowded that a lesion of the medulla may involve the motor tract on both sides, causing total bilateral paralysis—tetraplegia, usually due to thrombosis or to a small hæmorrhage. Or the arm on one side and the leg on the other may be involved—hemiplegia cruciata.

**Cerebellum.**—As “the head ganglion of the proprioceptive system” (Sherington) to this lesser brain converge the impulses of deep sensibility, and from it pass the impulses which control the tone of the muscles and their co-ordination when in action. The basis of our recent knowledge is in the exhaustive monograph of Luciani, whose conclusions have been confirmed and extended by Horsley and his pupils, Babinski, Thomas, and by the experience of the late war (Gordon Holmes).

In addition to its influence in maintaining equilibrium, the cerebellum has an important rôle in regulating and controlling voluntary movements. This is concerned with the muscular tone, the direction and measurement of movements, the maintenance of attitudes, and the control of co-ordinated movements. Hence disturbance of co-ordination, hypotonia, asthenia, ataxia (cerebellar) and volitional tremor result from diseased conditions. The disturbance may affect special functions. Ramsay Hunt described a condition under the designation *Dyssynergia cerebellaris progressiva*, or chronic progressive cerebellar tremor, in which there is a generalized volitional tremor which begins locally and gradually progresses. There is a progressive degeneration of the structures which control and regulate the muscular movements. When at rest and with the muscles relaxed the tremor ceases. Other symptoms of cerebellaris disease, such as vertigo, disturbance of equilibrium, nystagmus and seizures are absent.

**UNILATERAL LESIONS.**—As the functions of each lobe are homolateral, the symptoms are on the same side, and are negative not irritative in character. They may be grouped as follows (Gordon Holmes):

(1) *Disturbance of Muscle Tone.*—The limbs flop about in an unnatural way, and the muscles are soft and flabby. The hypotonia is so marked that with very little power the thigh can be flexed on the abdomen and the heel placed on the buttock. In walking the arm swings inertly, and if the fore-arms are held vertically, the wrist on the affected side falls passive in extreme flexion.

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(2) *Asthenia*, especially dwelt upon by Luciani, was a feature almost constant in the war cases. It is noted when the patient holds the arms outstretched or raises a weight, and is well shown by the dynamometer. The movements are slow, a delay in initiation and in relaxation. The affected limbs tire easily.

(3) *Ataria*.—In direction, force and range, the purposive movement errs, and with the eyes open. With the arm outstretched, asked to touch the nose with the index finger, he will bring it to the chin, and with undue force. Natural movements may be decomposed (Babinski), e.g., when asked to touch the knee with the heel, instead of flexing thigh and leg together, the hip is first flexed and then the knee. This *asynergia* is due to a lack of the proper association of agonists, antagonists and fixating muscles. The movements are ill measured (*dysmetria*), particularly quick movements, in both force and aim, and not along the shortest possible line. Tremor may occur in the moving limb, sometimes "intention" in character, or static, as in slight oscillations of the head when at rest; more characteristic is the tremor occurring in maintaining an attitude and involuntary movement, to which Luciani has given the name *Astasia*.

(4) *The Rebound Phenomenon*.—With elbows supported the patient pulls each hand towards his mouth against the resistance of the observer who holds the wrist. If let go suddenly, the hand on the affected side flies to the mouth often with great force, while the other is arrested almost immediately by the antagonists. This is a striking and valuable test.

(5) *Adiadokocinesis*.—In executing alternate movements as in rapid pronation and supination of the elbow, the homolateral limb moves more slowly, less regularly, and tires earlier, and there may be adventitious movements of the fingers.

(6) With *vertigo* the tendency is to fall towards the affected side, but the sensation of displacement may be of self or of external objects. It seems a more constant symptom in tumor than in injury.

(7) *The Pointing Test* (Bárány).—With closed eyes the patient is asked with his extended forefinger to touch the observer's finger held at some distance above the bed, and then as he brings the finger down to the bed and slowly up again the finger deviates outwardly.

(8) *Attitude and Gait*.—The head tends to be flexed towards the side of the lesion and rotated to the opposite side; and the body may be concave to the side of the lesion. On standing he is shaky and unsteady, and tends to fall towards the affected side, often with a feeling as if he were pulled over. The attitude may be very striking, the head and trunk inclined to the affected side, the spine concave to it, with the pelvis tilted, the shoulder lifted, the trunk rotated and held stiff. There is no Romberg sign. In walking he mistrusts the affected leg, which is usually rotated outwards, the foot may be dragged or raised unnaturally and brought to the floor with a flop. Stumbling towards the affected side, he makes efforts to control the tendency to fall. When asked to stop, he can not pull up suddenly. The arm on the affected side hangs inertly, without the normal swing.

(9) *Ocular Disturbances*.—In wounds there is early deviation of the eyes to the opposite side—or "skew deviation," the homolateral eye down and in, the other up and out. Fixation nystagmus is the rule in injury, and the oscilla-

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tions are slower and larger when the patient looks to the affected side. How far it is due to coexisting labyrinthine lesion is not determined, but Wilson and Pike claim that there are differences, and it is more enduring.

Among minor features to be mentioned are a slow, "sing-song" speech, the words are blurred, and the articulation nasal and the end syllables explosive. The homolateral reflexes may for a time be absent. As a rule the knee jerk is less brisk, and has a pendulum character. The superficial reflexes are not changed. Sensation in any form is unchanged.

*Bilateral lesions* show disturbances similar to those described above, but speech is more disturbed, the muscles of the trunk and neck are very hypotonic, and naturally when standing the maintenance of equilibrium is much more difficult. The features so characteristic of unilateral lesion are not essentially changed when the *vermis* is involved, unless perhaps the tremor is more marked. The effects of cortical and nuclear lesions do not appear to differ. The war experience does not support the view of special cortical localization, or of the existence of focal centres for movement in different directions (Bárány). The numerous clinical observations confirm Luciani's conclusions that *atonia*, *asthenia*, and *astasia* form a characteristic cerebellar triad.

## II. APHASIA

Under the general term aphasia—with agnosia and apraxia—is included the loss of the memories of the vocal, written, manual and other signs and symbols by which we communicate with our fellows and indicate our knowledge of the nature and use of things. As in all voluntary movements speech requires not only a motor but a sensory apparatus, and we have, as composing the speech mechanism, a sensory or receptive part as well as a motor or emissive part. These two parts are associated with the higher centres underlying the intellectual process, and are controlled by them.

The muscles which are used in the production of articulate speech are many and widely distributed; thus, the respiratory muscles of the larynx, the pharynx, the tongue, the lips, and those which move the jaws are all brought into play during speech. These muscles are active in other less complicated movements; for instance, respiration, crying, sucking, etc., and these comparatively simple movements are represented in the gray matter of the lower motor segment in the pons, medulla, and spinal cord. The association of neurones upon which these movements depend is made during fetal life, and is in good working order at the time of birth.

As the child's brain grows and takes control of the spinal centres through the medium of the pyramidal tracts, other more complex movements are developed and special neurones are set apart for this purpose. There is, then, a re-representation (Hughlings Jackson) of the finer movements of these muscles in the upper motor segment. They are localized in the central convolution about the lower part of the Rolandic fissure.

This group of movements, which are in part congenital and in part acquired during the early months of life, is that from which the delicate movements of articulate speech are developed. The structures upon which these movements depend make the *primary or elementary speech mechanism*.

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The cortical centres are in the lower third of the central convolution on both sides of the brain. They are bilaterally acting centres, and a lesion limited to either one should not produce marked or permanent defects in speech. This is true for the right side, but on the left Broca's convolution is so closely situated that it or its connecting fibres are usually injured at the same time, and aphasia results. The path from the cortical centres is made up of the motor fibres which go to the nuclei of the pons and medulla, and in the internal capsule is situated near the knee. As in the cortex, a unilateral lesion here causes only slight disturbances of speech due to difficult articulation, following weakness of the opposite side of the face and tongue. On the left side, if the lesion is so near the cortex as to involve the fibres which connect Broca's convolution with the primary speech mechanism, *sub-cortical motor aphasia* is produced. Bilateral lesions (usually in the internal capsule, but at times in the cortex) cause speechlessness, with paralysis of the muscles of articulation—pseudo-bulbar paralysis. To these speech defects Bastian gave the name *aphemia* and Marie, *anarthria*.

The lower segment of the primary speech mechanism is made up of the motor nuclei in the medulla, etc., and the peripheral nerves arising from them. Lesions here, if extensive enough—as, for instance, in progressive bulbar paralysis—may cause speechlessness—*anarthria* (Bastian); but usually they are more limited, giving various disturbances of articulation.

**The Auditory Speech Centre.**—As the child learns to speak there is developed in the cortex of the brain an association of centres which takes control of the primary speech mechanism. The child is constantly hearing objects called by names, and he learns to associate certain sounds with the look, feel, taste, etc., of certain things. When he hears such a sound he gets a more or less clear mental picture, or, in other words, he has developed certain auditory memories. These memories of the sounds of words are stored in what is called the *auditory speech centre*. This centre, which in the majority of people is the controlling speech centre, is situated on the left side in right-handed people and on the right side in those who are left-handed. The afferent impressions arising in the ears reach the transverse gyri of the temporal lobes, those from each ear going to both sides of the brain. From each of these primary auditory centres impulses are sent to the auditory speech centre in the temporal lobe of the left hemisphere. The exact location of this so-called centre is not accurately determined, but it is thought to occupy the first and perhaps part of the second temporal convolutions. Marie denies all speech centres, but places the cortical region, which has to do with the intellectual processes underlying language, rather vaguely in the left temporoparietal lobe. This he designates "*Wernicke's zone*," a lesion of which alone can produce aphasia. The child endeavors, and by repeated efforts learns, to make the sounds that he hears, and he first becomes able to repeat words, then to speak voluntarily. To do this, he has to learn certain very delicate movements, and so there is developed a special motor centre for speech in which these movements are localized.

**The Motor Speech Centre.**—This was placed by Broca, and those who immediately followed him, in the posterior part of the left third frontal convolution. It is around this—Broca's centre—that the discussion started by Marie has been most heated. Marie and his followers deny that this portion of the

brain has anything to do with speech, and insist that the so-called motor aphasia is merely a "combination of aphasia (of which they admit but one type, that due to lesions of Wernicke's zone) with anarthria." Anarthria they think of as a speech disturbance without any intellectual defect, due to a lesion of their lenticular zone, an ill-defined area in the centre of the brain.

Marie's position has been much discussed, and many excellent observers have come to the rescue of the old view which accepts Broca's convolution as the motor speech centre. The studies of cases of apraxia, which seem to have determined a centre in the left frontal lobe for certain purposive movements, as in the use of objects, gestures, etc., have lent support to the importance of Broca's convolution.

The motor speech centres and the corresponding area in the right brain are connected either directly by special motor fibres with the bulbar nuclei, or, as is more probable, indirectly, through the medium of the cortical centres of the primary speech mechanism in the lower part of the Rolandic region on both sides.

The speech centres are in close connection with the rest of the brain cortex, and in this way they take part in the general mental activities, of which, indeed, the speech processes form a large part. Some authors have assumed that the several sensory elements which go to make a concept are brought together in a special region of the brain, and here, as it were, united by a name. This is called "the centre for concepts," or "naming centre" (Broadbent), but most writers have followed Bastian in considering that the supposition of such a centre is unnecessary.

The mechanism which has been described is that which is developed in uneducated people and in children before they have learned to read and write, and is of primary importance in all speech processes. As the child learns to read he associates certain visual impressions with the speech memories he has acquired, and then adds to his concepts the visual memories of written or printed symbols. These memories are stored in the visual speech centre.

**The Visual Speech Centre.**—This is placed by nearly all authors in the angular and supramarginal convolutions on the left side, where it is believed visual impressions from both occipital lobes are combined in speech memories. Von Monakow denies such a special centre, but holds that visual speech memories are dependent upon the direct connection of the general visual centres in both occipital lobes with the speech sphere. That speech defects result from injury to the angular and supramarginal convolutions, he admits; but he thinks these are due to an interruption of fibre tracts which lie beneath and not to a destruction of a cortical centre. The distinction is, therefore, of more theoretical than practical importance. Marie includes this region in his Wernicke's zone.

In learning to write, the child develops certain delicate movements of the arm and hand, and thus acquires another method of externalizing his speech activities. Whether or not this requires the development of a separate writing centre, apart from the general Rolandic arm centre, or is brought about by an evolution of the latter through the medium of Broca's convolution, is a vexed question. Gordinier recorded a case of total agraphia, with no sensory or motor speech aphasia, in which a tumor occupying the foot of the second left frontal convolution was found at autopsy. *Agraphia* is a special form of

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**apraxia.** The movements of writing are learned under the influence of visual impressions in association with other speech memories, although there is a more direct path, which is used in copying unknown characters. Just as the movements of articulate speech are constantly under the control of auditory memories, so are the movements of writing regulated by visual memories; but in this case the other speech memories are of great importance.

With the development of the associations which underlie reading and writing, the speech mechanism may be said to be complete, although its activities are capable of practically endless extension, as when music or foreign languages are learned.

It will be seen that the cortical speech centres—the speech sphere of the French—occupy the part of the brain near the Sylvian fissure, and that they all receive their blood from the Sylvian artery. Speaking broadly, the posterior part of this region is sensory and the anterior is motor. The sensory areas are near the optic radiation and the motor are near the general motor tracts, and so, with lesions of the posterior part, hemianopia is apt to be associated with the speech disturbance while hemiplegia occurs with disease of the anterior area. These associations often help to distinguish a sensory from a motor aphasia, but each type has special characteristics.

**Auditory Aphasia.**—Most people in mentally recalling words do so by means of their auditory speech memories—i. e., they think of the sound of the words, and, in voluntary speech, it is probable that the will acts on the motor centre indirectly through the auditory centre. This centre is also necessary for reading in such persons. There are persons, however, in whom the mental processes are carried on by visual memories, and in these “visuals” the visual speech centres takes the predominant place in speech usually occupied by the auditory centres.

Complete abolition of all the auditory speech memories by destruction of the first temporal convolution causes the most extensive disturbances of speech. Such a person is unable to comprehend speech, either spoken or printed. Voluntary speech is much disturbed, and although at first he may talk, his speech is nothing but a jargon of misplaced words, and he soon becomes speechless. Writing is also lost, and he can neither repeat words nor write at dictation. He may be able to copy.

Lesions are often only partial, and the resultant disturbance may be simply a difficulty in speech due to the loss of nouns or to the transposition of words (paraphasia), the writing showing the same defect. The patient usually understands what he hears and reads, and can repeat words and write at dictation. Bastian called this condition “*amnesia verbalis*.” It may be so pronounced that voluntary speech and writing are nearly lost, even when the auditory memories can still be aroused by new afferent impressions and he is able to understand what is said to him and what he reads. He can usually repeat and read aloud.

The afferent paths, which reach the auditory speech centre from the two primary auditory centres, may be destroyed. A lesion to do this must be in the white matter beneath the first temporal convolution on the left side. Such a lesion blocks all auditory impressions coming to the centre, and the patient is not able to understand anything said to him, cannot repeat words or write from dictation. As the cortical centres are not disturbed, and the audi-

## APHASIA

ory speech memories are still present, there is no disturbance of voluntary speech or writing, and the patient can read perfectly. This is pure word-deafness or subcortical sensory aphasia.

**Visual Aphasia.**—Destruction of the visual centre in the angular and supramarginal convolutions causes a loss of the visual speech memories, and the patient is unable to read printed or written characters. He is unable to write—agraphia—and he can not copy. His understanding of spoken words is good, and voluntary speech is normal or only slightly paraphasic.

A subcortical lesion involving the afferent fibres going to the visual speech centre causes pure word-blindness (subcortical alexia)—i. e., there is inability to understand written or printed words. Voluntary speech and writing are good. The patient can not read his own writing except by aid of muscle-sense impression, in retracing the letters, either voluntarily or passively. Associated with this is always hemianopia.

Word-deafness and word-blindness are often combined, and at times it is not only the tracts that connect the primary auditory and visual centres with the speech spheres, but also those which associate them with the other sensory centres in the formation of concepts, that are diseased. In this case the patient has lost not only his auditory and visual speech memories, but also all of his memories which have to do with hearing and sight. He has mind-deafness and mind-blindness—i. e., he is unable to recognize objects when he hears or when he sees them. Further, there may be a dissociation of all the sensory centres from each other or from the higher psychical centre, which is practically the same thing, in which case the patient is entirely unable to recognize objects or use them properly—i. e., he has sensory apraxia or agnosia.

**Motor Aphasia.**—Lesions of the motor speech zone, possibly in rare cases of Broca's convolution alone, more commonly of a wider area, cause loss of the power of speech. The patient may be absolutely dumb, or he may have retained one or two words or phrases, which is believed to be due to the activity of the corresponding region of the right brain. He will make no effort to repeat words. His mind is comparatively clear, and he understands what is said to him, but reads poorly. He has not a clear mental picture of words. This is tested by asking him to squeeze the observer's hand or to make expiratory efforts as many times as there are syllables in a well-known name.

Voluntary writing is usually lost in cortical motor aphasia, and many authors believe that writing movements are controlled from this centre. Others, who believe that there is a special writing centre, contend that a lesion strictly limited to the motor speech centre would not cause agraphia, and cite cases which seem to support their view. If there is much disturbance of internal speech, writing must be impaired.

*Subcortical motor aphasia* is described as due to the destruction of the fibres which join Broca's convolution to the primary speech mechanism. Lesions which have produced this type of aphasia have been in the white matter of the left hemisphere near Broca's convolution. These would be within Marie's lenticular zone. There is complete loss of the power of speech without any disturbance of internal speech. The patient's mental processes are not disturbed, and he can write perfectly if the hand is not paralyzed.

Cases of aphasia are rarely simple, and it is often impossible to classify them accurately. The problems involved are exceedingly complicated, and

the student must not for a moment suppose that cases are as straightforward as the various diagrams at first sight appear to indicate. A majority are very complex, but with patience the diagnosis of the different varieties can often be worked out. The following tests should be applied, after the presence or absence of paralysis has been determined and whether the patient is right- or left-handed: (1) The power of recognizing the nature, uses, and relations of objects—i. e., whether agnosia and apraxia are present or not; (2) the power to recall the name of familiar objects seen, smelled, or tasted, or of a sound when heard, or of an object touched; (3) the power to understand spoken words; (4) the capability of understanding printed or written language; (5) the power of appreciating and understanding music; (6) the power of voluntary speech—in this it is to be noted particularly whether he misplaces words or not; (7) the power of reading aloud and of understanding what he reads; (8) the power to write voluntarily and of reading what he has written; (9) the power to copy; (10) the power to write at dictation; and (11) the power of repeating words.

The *medico-legal aspects* of aphasia are of great importance. No general principle can be laid down, but each case must be considered on its merits. Langdon, in reviewing the whole question, concludes: "Sanity established, any legal document should be recognized when it can be proved that the person making it can understand fully its nature by any receptive channel (viz., hearing, vision, or muscular sense), and can, in addition, express assent or dissent with certainty to proper witnesses, whether this expression be by spoken speech, written speech, or pantomime."

*Prognosis.*—In young persons the outlook is good, and the speech is gradually restored apparently by the development of other portions of the brain. The opposite hemisphere often takes part in this. In adults the condition is less hopeful, particularly in complete motor aphasia with right hemiplegia. The patient may remain speechless, though capable of understanding everything, and attempts at re-education may be futile. Partial recovery may occur, and the patient may be able to talk, but misplaces words. In sensory aphasia the condition may be only transient, and the different forms rarely persist alone without impairment of the powers of expression.

The *education* of an aphasic person requires the greatest care and patience, particularly if, as often happens, he is emotional and irritable. It is best to begin by the use of detached letters, and advance slowly to words of only one syllable. Children often make rapid progress, but in adults failure is only too frequent, even after the most painstaking efforts. In right-sided hemiplegia with aphasia the patient may be taught to write with the left hand.

### III. AFFECTIONS OF THE BLOOD-VESSELS

#### I. ARTERIO-SCLEROSIS—CEREBRAL FEATURES

(1) **Transient Paralysis.**—With high blood pressure and sclerotic vessels attacks of aphasia, monoplegia and hemiplegia occur, with the following characters: they are transient, leave no permanent damage, and recur. Pure cases of motor aphasia are met with—a twelve to twenty-four hour inability

to speak, without any mental disturbance. Monoplegia of the arm alone, or with the face, is more common than hemiplegia. A patient may have scores of attacks over many years. They are often associated with increased blood pressure and headache. Numbness and tingling or twitching of the angle of the mouth or the hand may precede an attack. One patient had transient hemianopia. Sudden paraplegia may come on and last part of a day. Coming down the gangway of a steamer, a friend who had had many attacks of monoplegia suddenly lost power in the legs, and had to be carried. He could walk next day. Another dropped in the street, and when seen twelve hours later, the paralysis was just disappearing and the reflexes obtainable. These are not attacks of intermittent claudication. The possible influence of increased concentration or viscosity of the blood, as from vomiting or diarrhoea, should be considered. It may also have an influence in apoplexy. It is important to prevent dehydration in old persons. Inman has emphasized the importance of *slowing* of the cerebral circulation in explaining the temporary disturbances in arterio-sclerosis and as also favoring thrombosis. He warns against fatigue and suggests exercises, with the patient lying down, especially of the abdominal muscles to aid the splanchnic circulation.

(2) **Convulsions** may occur with the above attacks or independently. The attack rarely has the graded features of true epilepsy, but there are widespread clonic movements, with unconsciousness for a few minutes to an hour. There may be daily attacks and transient paralysis may follow on aphasia. The general condition may remain good and the mental state undamaged.

(3) **Psychical Changes.**—Following a convulsion, the patient may be dazed and “not himself” for some hours. A remarkable feature in many cases is the retention of exceptional mental vigor. A transient mental outburst may replace, as it were, the motor attack. One subject of innumerable monoplegias would waken at night, light the candle, stamp about the room, tear up books and papers, all the time talking to himself. He would know nothing about it in the morning. Similar outbursts occurred in the day. Or a transient cloud may pass over the mind before the onset of hemiplegia. Returning from a game of golf, a man did not know his house or recognize his wife and surroundings. After a good night’s rest he woke with weakness of the right side and confusion of speech which had gone by the evening.

As the disease progresses, the mental state may fail, but in contradistinction to the presenile and senile types of dementia, many of these patients keep a clear mind to the end, and there are none of the features of Binswanger’s *dementia presenilis* or of Alzheimer’s disease. An explanation of these attacks is not easy. Their frequency and the rapid restoration of function rule out destructive lesions. Possibly they are due to spasm of the arteries and a temporary ischæmia, a view strongly supported by similar attacks in Raynaud’s disease, or to a temporary œdema.

Clinically there are three groups of cases: (1) The arterio-sclerosis of middle-aged men; (2) the senile form; and (3) special presenile forms.

(1) **ARTERIO-SCLEROSIS** (see p. 860 under diseases of the arteries).

(2) **SENILE ARTERIO-SCLEROSIS.**—Old age is largely a question of the blood vessels, but the wear and tear of life affects different parts in different persons. With the progressive weakening of the mental powers as age advances, widespread changes in the arteries, both basal and cortical, are found. Often

it is not a question of the petrol-tank—the blood supply—but the whole machine is worn out. A real mental vigor may exist with advanced arteriosclerosis. A man of sixty in full practice at the bar died suddenly of angina pectoris. The basal arteries were pipe-stems, and the smaller cortical vessels creaked under the knife!

In a normal old age the convolutions waste, the pigment granules and the lime-salts increase, the meninges become cloudy, the cortical arteries thicken, the glia in the gray matter increases particularly about the smaller vessels, and there are the areas of atrophy, as described by Marie, Peck and others. With these organic changes the mental grip fails, the memory weakens, the emotions are less under control, and year by year in a slow process of devolution the last stage of all is reached, second childhood—babyhood rather—as the man ends as he began, with only a vegetative system.

This happy, normal process with "mild gradations of decay," recognized by all except the senile himself, bears out Plato's dictum that "old age is an easy death." But it may be far otherwise, and "the evening of life may be a stormy and unhappy period." The peculiarities of the individual become more marked and to an unpleasant degree; he becomes egotistical, emotional and suspicious, or careless in the minor proprieties of life and intensely selfish. The most pathetic of martyrdoms are the miseries endured by children in the unrequited, unappreciated devotion to an irritable, egotistical, self-centred senile parent. But the pity of it is that the worst troubles may not be intensification of any personal peculiarities, but terrible perversions of character of a distressing nature. The man of active useful life may be depressed to distraction by the thoughts of the failure he has been; the godly man is worried over his lost soul; the moral teacher and saintly soul may become a lecher; or the loving affectionate husband a brutal tyrant.

3. SPECIAL TYPES.—While in normal old age there is nothing local, on the other hand, the senility may be chiefly local and affect the brain at a comparatively early age. The changes are usually those of normal old age, and associated with loss of judgment, emotional perversions, and progressive mental impairment. The cardio-vascular and renal conditions play an important rôle in these cases (Southard). Various forms have been described.—the *presbyophrenia* of Wernicke—characterized by "marked disturbances of the recording faculty, with retention for a long time of orderly thought and judgment . . . and tendency to confabulation" (Barker). Binswanger's *dementia presenilis* begins between the ages of 40 and 50, with loss of memory, apathy, etc., without syphilis or the somatic feature of *general paresis*. *Alzheimer's* disease is a slow dementia with focal symptoms, aphasia and apraxia, and in addition to the regressive changes in the vessels and glia, a peculiar condition of the neuro-fibrils. Southard and Alford called attention to a group of senile dementias (11 of 12 cases specially studied) of obscure etiology, which do not come in these types as the vessels are not sclerotic and the convolutions are not atrophied.

## II. HYPERÆMIA AND ANÆMIA

Less and less stress is now laid on these conditions. The symptoms usually referred to active hyperæmia in the infectious diseases, or in association with

hypertrophy of the heart accompanying disease of the kidney, are due to the action of toxic agents rather than to changes in the circulation.

*Anæmia.*—The anatomical condition of the brain is very striking. The membranes are pale, only the large veins are full, the small vessels over the gyri are empty, and an unusual amount of cerebro-spinal fluid is present. On section both the gray and white matter look extremely pale and the cut surface is moist. Very few *puncta vasculosa* are seen.

*Symptoms.*—The effects of sudden anæmia of the brain are well illustrated by the ordinary fainting fit. When the symptoms are the result of hæmorrhage, there are drowsiness and giddiness; flashes of light, dark spots before the eyes, and noises in the ears; the respiration becomes hurried; the skin is cool and covered with sweat; the pupils are dilated, there may be vomiting, headache, or delirium, and gradually, if the bleeding continues, consciousness is lost and death may occur with convulsions. In the more chronic forms, such as result from anæmia, a condition of irritable weakness results. Mental effort is difficult, the slightest irritation is followed by undue excitement, the patient complains of giddiness and noises in the ears, or there may be hallucinations or delirium. These symptoms occur in an extreme grade as a result of prolonged starvation, and a similar condition is seen in certain cases of arterio-sclerosis, when the brain is poorly nourished.

An interesting set of symptoms, to which the term *hydrencephaloid* was applied by Marshall Hall, occurs in the anæmia and debility produced by prolonged diarrhoea in children. The child is in a semi-comatose condition with the eyes open, the pupils contracted, and the fontanelle depressed. In the earlier period there may be convulsions. The coma may gradually deepen, the pupils become dilated, and there may be strabismus and even reaction of the head, symptoms which closely simulate those of basilar meningitis.

### III. ŒDEMA OF THE BRAIN

Whether it occurs as a clinical entity is doubtful. As a secondary process it occurs under the following conditions: In general atrophy of the convolutions, in which case the œdema is represented by an increase in the cerebro-spinal fluid and in that of the meshes of the pia. In extreme venous dilatation from obstruction, as in mitral stenosis or tumors, there may be congestive œdema, in which, in addition to great filling of the blood-vessels, the substance of the brain itself is unusually moist. The most acute œdema is a local process found around tumors and abscesses. The symptoms of compression following concussion or contusion, as shown by Cannon, are frequently attributable to cerebral œdema due to change in osmotic pressure. An intense infiltration, local or general, may occur in nephritis, and to it certain of the uræmic symptoms may be due. There is a form, termed "wet brain," found in alcoholics.

*Anatomical Changes.*—These are not unlike those of anæmia. When the œdema follows progressive atrophy, the fluid is chiefly within and beneath the membranes. The brain substance is anæmic and moist and has a wet, glistening appearance, which is very characteristic. In some instances the œdema is more intense and local, and the brain substance may look infiltrated with fluid. The amount of fluid in the ventricles is usually increased.

**Symptoms.**—The symptoms are in great part those of lessened blood flow and are not well defined. Some of the cerebral features of uræmia may depend upon it. Cases have been reported in which unilateral convulsions or paralysis have occurred in connection with chronic nephritis, and in which the condition appeared to be associated with cedema of the brain. The older writers laid great stress upon an apoplexia serosa, which may really have been a general cedema of the brain. Some of the cases of transient paralysis or aphasia may be caused by cedema.

**Treatment.**—Associated conditions require the proper treatment. Lumbar puncture is indicated and should be repeated when necessary. The bowels should be freely moved by salines. In the "wet brain" of alcoholism ergot in full doses is useful.

#### IV. CEREBRAL HÆMORRHAGE

The bleeding may come from branches of either of the two great groups of cerebral vessels—the *basal*, comprising the circle of Willis and the central arteries passing from it and from the first portion of the cerebral arteries, or the *cortical group*, the anterior, middle, and the posterior cerebral vessels. In a majority of the cases the hæmorrhage is from the central branches, more particularly from those which are given off by the middle cerebral arteries in the anterior perforated spaces, and which supply the corpora striata and internal capsules. One of the largest of these branches which passes to the third division of the lenticular nucleus and to the anterior part of the internal capsule, the lenticulo-striate artery, is so frequently involved in hæmorrhage that it was called by Charcot *the artery of cerebral hæmorrhage*. Hæmorrhages from this and from the lenticulo-thalamic artery include more than 60 per cent. of all cerebral hæmorrhages. The bleeding may be into the substance of the brain, to which alone the term cerebral apoplexy is applied, or into the membranes, in which case it is termed meningeal hæmorrhage; both are usually included under the terms intracranial or cerebral hæmorrhage.

**Etiology.**—High blood pressure and arterial disease in persons over forty years of age are the main factors.

**AGE.**—After thirty the liability increases with each decade. It may be congenital as in the child of a woman dead of typhoid fever at the Johns Hopkins Hospital. It occasionally occurs in children from rupture of a small aneurism, but before the age of thirty it is very uncommon. In an analysis of the United States Census Report. H. M. Thomas found the increase common in the 7th and 8th decades. Of 154 cases at St. Bartholomew's Hospital traceable to arterial changes there was no case under thirty; the maximum for both sexes was at the fifty-sixth year. After sixty the numbers appear to decline, but if "due correction is made for the age-distribution of a population, the liability of the individual to this form of death increases steadily up to old age" (F. W. Andrewes). Before the fifth decade hæmorrhage is rare; then in the fifth and sixth decades cases progressively increase in number.

**SEX.**—There is a marked preponderance of males.

**RACE.**—In the United States the death rate from apoplexy in the Report of 1920 was 809 per million population. In England and Wales in 1922 the

deaths from apoplexy were 706 per million living. Both apoplexy and paralysis seem to be much more prevalent among the negroes.

**HEREDITY.**—Formerly thought to be a very important factor, heredity influences the incidence in rendering members of families in which the blood vessels degenerate early more liable to cerebral hæmorrhage. What was known as the apoplectic habitus, or build, is still spoken of, by which we mean a stout, plethoric person of medium size with a short neck.

**SPECIAL FACTORS.**—Individuals with progressive renal disease and consecutive arterio-sclerosis and hypertrophy of the heart are particularly liable to cerebral hæmorrhage. Alcohol, immoderate eating, prolonged muscular exertion, syphilis, and gout are antecedents in many cases. Endocarditis may lead indirectly to apoplexy by causing embolism and aneurism of the vessels of the brain. Hæmorrhage may occur with new growths. Cerebral hæmorrhage occurs occasionally in the specific fevers and with profound alterations of the blood as in leukaemia.

The actual exciting cause is not always evident. The attacks may be sudden without any preliminary symptoms. In other instances straining efforts or overaction of the heart may cause a rupture. Some cases occur during sleep or follow slight trauma. The records of University College Hospital analyzed by Ernest Jones indicate that in none of 123 cases did the attack come on through excessive bodily effort.

**Morbid Anatomy.**—**DIRECT CHANGES.**—The lesions are almost invariably in the cerebral arteries, in which the following changes may lead to it:

(a) The rupture of miliary aneurisms is a common cause of cerebral hæmorrhage. They occur most frequently on the central arteries, but also on the smaller branches of the cortical vessels. On section of the brain they may be seen as localized, small dark bodies, about the size of a pin's head. Sometimes they are seen in numbers upon the arteries when withdrawn from the anterior perforated spaces. In apoplexy after the fortieth year if sought for they are rarely missed.

(b) Aneurism of the branches of the circle of Willis. These are by no means uncommon, and will be considered subsequently.

(c) Endarteritis and periarteritis in the cerebral vessels most commonly lead to hæmorrhage by the production of aneurisms, either miliary or coarse. There are instances in which the most careful search fails to reveal anything but diffuse degeneration of the cerebral vessels.

(d) Whether hæmorrhage ever occurs by *diapedesis* without actual rupture is doubtful. Possibly it does in purpura.

(e) In persons over sixty the hemiplegia may depend upon small areas of softening in the gray matter—the *lacune* of Marie—varying in size from a pin's head to a pea or a small bean, grayish red in tint. The lenticular nucleus is particularly apt to be involved. The blood-vessels are always diseased.

The hæmorrhage may be meningeal, cerebral, or intraventricular.

**Meningeal hæmorrhage** may be outside the dura, between dura and arachnoid, or between the arachnoid and the pia mater. The following are the chief causes: Fracture of the skull, in which case the blood usually comes from the lacerated meningeal vessels, sometimes from the torn sinuses. In these cases the blood is usually outside the dura or between it and the arachnoid. The next most frequent cause is rupture of aneurisms on the larger

cerebral vessels. The blood is usually subarachnoid. An intracerebral hæmorrhage may burst into the meninges. A special form of meningeal hæmorrhage is found in the new-born, associated with injury during birth. And lastly, meningeal hæmorrhage may occur in the constitutional diseases and fevers. The blood may be in a large quantity at the base and may extend into the cord or upon the cortex. Owing to the greater frequency of aneurisms in the middle cerebral vessels, the Sylvian fissures are often distended with blood.

*Intracerebral hæmorrhage* is most frequent in the neighborhood of the corpus striatum, particularly toward the outer section of the lenticular nucleus. The hæmorrhage may be small and limited to the lenticular body, the thalamus, and the internal capsule, or it may extend to the insula. Hæmorrhages confined to the white matter—the centrum semiovale—are rare. Localized bleeding may occur in the crura or in the pons. Hæmorrhage into the cerebellum usually comes from the superior cerebellar artery. The extravasation may be limited to the substance or may rupture into the fourth ventricle.

*Ventricular Hæmorrhage.*—This is rarely primary, coming from the vessels of the plexuses or of the walls. More often it is secondary, following hæmorrhage into the cerebral substance. It is not infrequent in early life and may occur during birth. Of 94 cases collected by Edward Sanders, 7 occurred during the first year, and 14 under the twentieth year. In adults it is almost always caused by rupture of a vessel in the neighborhood of the caudate nucleus. The blood may be found in one ventricle only, but more commonly it is in both lateral ventricles, and may pass into the third ventricle and through the aqueduct of Sylvius into the fourth ventricle, forming a complete mould in blood of the ventricular system. In these cases the clinical picture may be that of "*apoplexie foudroyante*."

*Multiple Hæmorrhages.*—Of 128 non-traumatic cases at the Cook County Hospital there were 28 with discrete multiple hæmorrhages. The most common form is hæmorrhage into the basal ganglia and into the pons; the next, bilateral basal hæmorrhage. In the brain compression following hæmorrhage, the blood pressure rises; this increased intracranial tension is doubtless the cause of rupture in other vessels weakened by disease. The pontine arteries seem specially susceptible, as the small terminal vessels come off at right angles to a very large trunk (Phyllis Greenacre).

**SUBSEQUENT CHANGES.**—If the hæmorrhage is of any extent there is compression of the brain, which, with a secondary œdema, may play a part in causing coma. The blood gradually changes in color, and ultimately the hæmoglobin is converted into hæmatoidin. Inflammation occurs about the area, limiting it, and ultimately a definite wall may be produced, inclosing a cyst with fluid contents. In other instances a cyst is not formed, but the connective tissue proliferates and leaves a pigmented scar. In meningeal hæmorrhage the effused blood may be gradually absorbed and leave only a staining of the membranes. In other cases, particularly in infants, when the effusion is cortical and abundant, there may be localized wasting of the convolutions and the production of a cyst in the meninges. Possibly porencephaly may arise in this way. Secondary degeneration follows, involving various tracts according to the location of the hæmorrhage and the damage done by it to nerve cells or their medullated axones.

**Symptoms.**—**PRIMARY.**—Premonitory indications are rare. As a rule, the patient is seized while in full health or in the performance of some every day action, occasionally an action requiring strain or extra exertion. There may be headache, sensations of numbness or tingling or pains in the limbs, or even choreiform movements in the muscles of the opposite side, the so-called prehemiplegic chorea. In other cases temporary disturbances of vision and of associated movements of the eye-muscles have been noted, but none of the prodromata of apoplexy (the so-called "warnings") are characteristic. Transient aphasia or monoplegia may precede the attack. The onset may be with sudden loss of consciousness and complete relaxation of the extremities. In such instances the name *apoplectic stroke* is particularly appropriate. In other cases it is more gradual and the loss of consciousness may not occur for a few minutes after the patient has fallen, or after the paralysis of the limbs is manifest. In the typical apoplectic attack the condition is as follows: There is deep unconsciousness; the patient can not be roused. The face is injected, sometimes cyanotic, or of an ashen gray hue. The pupils vary; usually they are dilated, sometimes unequal, and always, in deep coma, inactive. If the hæmorrhage be so located that it can irritate the nucleus of the third nerve the pupils are contracted (hæmorrhages into the pons or ventricles). The respirations are slow, noisy, and accompanied with stertor. Sometimes Cheyne-Stokes rhythm may be present. The chest movements on the paralyzed side may be restricted, in rare instances on the opposite side. The cheeks are often blown out during expiration, with spluttering of the lips. The pulse is usually full, slow, and of increased tension. The temperature may be normal, but is often subnormal, and, as in a case reported by Bastian, may sink below 95°. In basal hæmorrhage the temperature, on the other hand, may be high. The urine and faeces are usually passed involuntarily. Convulsions are not common. It may be difficult to decide whether the condition is apoplexy associated with hemiplegia or sudden coma from other causes. An indication of hemiplegia may be discovered in the difference in the tonus of the muscles on the two sides. If the arm or the leg is lifted, it drops "dead" on the affected side, while on the other it falls more slowly. The lack of muscular tone of the paralyzed limb may be determined by inspection; the muscle mass of the thigh acts like a semi-fluid sac and takes the shape determined by gravity. In a patient lying or sitting on a firm support, the thigh of the paralyzed limb is broadened or flattened, while that on the normal side has a more rounded contour. Rigidity may be present, especially in some cases of middle meningeal hæmorrhage. In watching the movements of the facial muscles in the stertorous respiration it will be seen that on the paralyzed side the relaxation permits the cheek to be blown out in a more marked manner. The head and eyes may be turned to one side—conjugate deviation. Sudden death is exceptionally rare and death hardly ever occurs under some hours.

In other cases, in which the onset is not so abrupt, the patient may not lose consciousness, but in the course of a few hours there is loss of power, unconsciousness comes on gradually, and deepens into profound coma—in-gravescent apoplexy. The attack may occur during sleep; the patient may be found unconscious or wakes to find that the power is lost on one side. Small hæmorrhages in the territory of the central arteries may cause hemi-

plegia without loss of consciousness. In old persons the hemiplegia may be slight and follow a transient loss of consciousness, and is usually most marked in the leg. It is associated with other senile changes. This form is often due to the presence of lacunar softening.

Usually within forty-eight hours after the onset, sometimes within from two to six hours, there are febrile reaction and some constitutional disturbance associated with inflammatory changes about the hæmorrhage and absorption of the blood. The period of inflammatory reaction may continue for from one week to two months. The patient may die in this reaction, or, if consciousness has been regained, there may be delirium or recurrence of the coma. \* At this period the so-called early rigidity may develop in the paralyzed limbs and trophic changes occur, such as sloughing or the formation of vesicles. The most serious of these is the sloughing eschar of the lower part of the back, or on the paralyzed side, which may appear within forty-eight hours of the onset and is usually of grave significance. The common congestion at the bases of the lungs is regarded by some as a trophic change.

*Conjugate Deviation.*—In a flaccid paralysis the eyes and sometimes the head may be turned away from the paralyzed side, that is, the eyes look toward the cerebral lesion. This is almost the rule in hemiplegia. When, however, convulsions or spasm occur or the state of so-called early rigidity, the conjugate deviation of the head and eyes may be in the opposite direction; that is to say, the eyes look away from the lesion and the head is rotated toward the convulsed side.

*Hemiplegia.*—When consciousness is restored and the patient improves, a unilateral paralysis may persist due to the destruction of the motor area or the pyramidal tract in any part of its course. Hemiplegia is complete when it involves face, arm, and leg, or partial when it involves only one or other of these parts. This may be the result of a lesion (a) of the motor cortex; (b) of the pyramidal fibres in the corona radiata and in the internal capsule; (c) of a lesion in the cerebral peduncle; or (d) in the pons Varolii. The situation of the lesions and their effects are given in Fig. 22. Vascular lesions are perhaps the most common cause, but tumors and spots of softening may also induce it. The special details of the hemiplegia may be considered. The face (except in lesions in the lower part of the pons) is involved on the same side as the arm and leg. This results from the fact that the facial muscles stand in precisely the same relation to the cortical centres as those of the arm and leg, the fibres of the upper motor segment of the facial nerve from the cortex decussating just as do those of the nerves of the limbs. The signs of the facial paralysis are usually well marked. There may be a slight difficulty in elevating the eyebrows or in closing the eye on the paralyzed side, or in rare cases, the facial paralysis is complete, but the movements may be present with emotion, as laughing or crying. The facial paralysis is partial, involving only the lower portion of the nerve, so that the orbicularis oculi and the frontalis muscles are much less involved than the lower branch. The hypoglossal nerve also is involved. In consequence, the patient can not put out the tongue straight, but it deviates toward the paralyzed side, inasmuch as the genio-hyo-glossus of the sound side is unopposed. In a few cases the protrusion is toward the side of the lesion, a

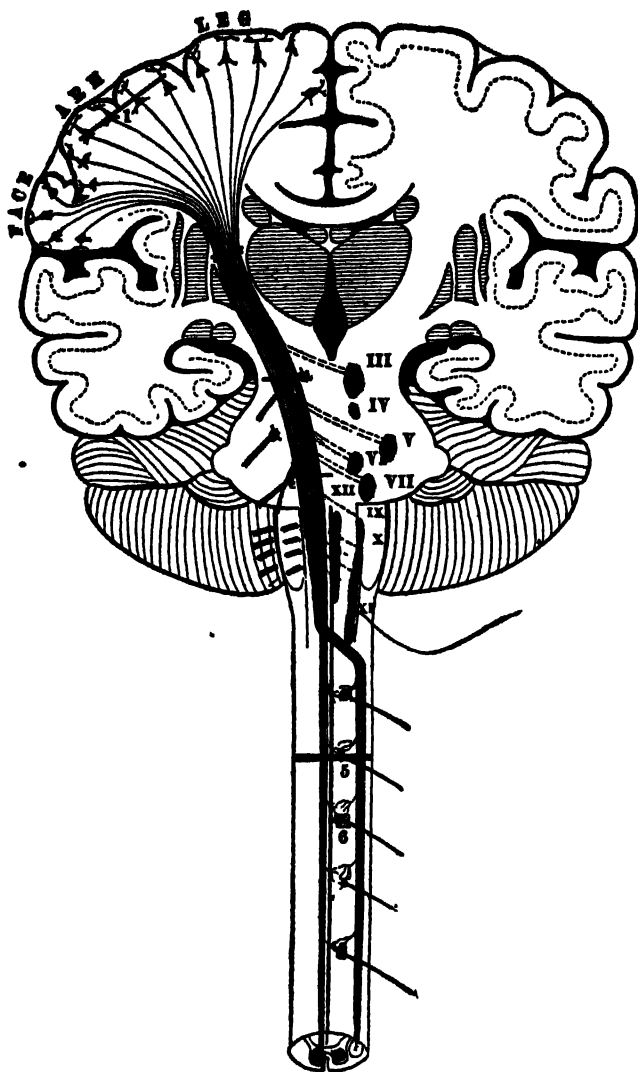


FIG. 22.—DIAGRAM OF MOTOR PATH FROM LEFT BRAIN.

The upper segment is black, the lower red. The nuclei of the motor cerebral nerves are shown on the right side, on the left side the cerebral nerves of that side are indicated. A lesion at 1 would cause upper segment paralysis in the arm of the opposite side—cerebral monoplegia; at 2, upper segment paralysis of the whole opposite side of the body—hemiplegia; at 3 (in the crus), upper segment paralysis of the opposite face, arm, and leg, and lower segment paralysis of the eye-muscles on the same side—crossed paralysis; at 4 (in the lower part of the pons), upper segment paralysis of the opposite arm and leg, and lower segment paralysis of the face and external rectus on the same side—crossed paralysis; at 5, upper segment paralysis of all muscles represented below lesion, and lower segment paralysis of muscles represented at level of lesion—spinal paraplegia; at 6, lower segment paralysis of muscles localized at seat of lesion—anterior poliomyelitis. (Van Gehuchten, modified.)

## DISEASES OF THE NERVOUS SYSTEM

fact not easily explained. With right hemiplegia there may be aphasia. Even without marked aphasia difficulty and slowness in speaking are common.

The arm is, as a rule, more completely paralyzed than the leg. The loss of power may be absolute or partial. In severe cases it is at first complete. In others, when the paralysis in the face and arm is complete, that of the leg is only partial. The face and arm may alone be paralyzed, while the leg escapes. Less commonly the leg is more affected than the arm, and the face may be only slightly involved.

Certain muscles escape in hemiplegia, particularly those associated in symmetrical movements, as those of the thorax and abdomen, which Broadbent explains by supposing that as the spinal nuclei controlling these movements on both sides constantly act together they may, by means of this intimate connection, be stimulated by impulses coming from only one side of the brain. Hughlings Jackson pointed out that in quiet respiration the muscles on the paralyzed side act more strongly than the corresponding muscles, but that in forced respiration the reverse condition is true.

The degree of permanent paralysis after a hemiplegic attack varies greatly. When the restitution is partial, it is always certain groups of muscles which recover rather than others. Thus in the leg the residual paralysis concerns the flexors of the leg and the dorsal flexors of the foot—i. e., the muscles which are active in the second period of walking, shortening the leg, and bringing it forward while it swings. The muscles which lift the body when the foot rests upon the ground, those used in the first period of walking, include the extensors of the leg and the plantar flexors of the foot. These "lengtheners" of the leg often recover almost completely when the paralysis is due to lesions of the pyramidal tract. In the arms the residual paralysis usually affects the muscle groups which oppose the thumb, those which rotate the arm outward, and the openers of the hand. As a rule, there is at first no wasting of the paralyzed limbs.

*Crossed Hemiplegia.*—A paralysis in which there is loss of function in a cerebral nerve on one side with loss of power (or of sensation) on the opposite side of the body is called a crossed or alternate hemiplegia. It is met with in lesions, commonly hemorrhage, in the crus, the pons, and the medulla.

(a) *Crus.*—The bleeding may extend from vessels supplying the corpus striatum, internal capsule, and optic thalamus, or the hemorrhage may be primarily in the crus. In the classical case of Weber, on section of the lower part of the left crus, an oblong clot 15 mm. in length lay just below the medial and inferior surface. The characteristic features of a lesion in this locality are paralysis of arm, face, and leg of the opposite side, and third nerve paralysis of the same side—the syndrome of Weber or Weber-Gubler. Sensory changes may also be present. Hemorrhage into the tegmentum is not necessarily associated with hemiplegia, but there may be incomplete paralysis of the oculo-motor nerve, with disturbance of sensation and ataxia on the opposite side. The optic tract or the lateral geniculate body lying on the lateral side of the crus may be compressed, with resulting hemianopia.

(b) *Pons and Medulla.*—Lesions may involve the pyramidal tract and one or more of the cerebral nerves. If at the lower aspect of the pons, the facial nerve may be involved, causing paralysis of the face on the same side and hemiplegia on the opposite side. The sixth nerve is also usually involved,

## AFFECTIONS OF THE BLOOD-VESSELS

causing paralysis of the external rectus. The fifth nerve may be involved, with the fillet (the sensory tract), causing loss of sensation in the area of distribution of the fifth on the same side as the lesion and loss of sensation on the opposite side of the body. The sensory disturbance here is apt to be dissociated, of the syringomyelic type, affecting particularly the sense of pain and temperature.

*Sensory Disturbances from Cerebral Hæmorrhage.*—These are variable. Hemianæsthesia may coexist with hemiplegia, but in many instances there is only slight numbing of sensation. When marked, it is usually the result of a lesion in the internal capsule involving the retrolenticular portion of the posterior limb. In a study of sensory localization Dana found that anæsthesia of organic cortical origin was always limited or more pronounced in certain parts, as the face, arm, or leg, and was generally incomplete. Total anæsthesia was either of functional or subcortical origin. Marked anæsthesia was much more common in softening than in hæmorrhage. Complete hemianæsthesia is rare in hæmorrhage. Disturbance of the special senses is not common. Hemianopia may exist on the same side as the paralysis, and there may be diminution in the acuteness of hearing, taste, and smell. Homonymous hemianopia of the halves of the visual fields opposite to the lesion is frequent shortly after the onset, though often overlooked (Gowers).

Psychic disturbances, variable in nature and degree, may result from cerebral hæmorrhage.

*Reflexes.*—With deep coma due to increased intracranial pressure the knee jerks and abdominal reflexes may be absent on both sides with a bilateral extensor plantar reflex. Later they may return on the non-hemiplegic side. On the hemiplegic side the lost reflexes may return only after weeks. As to the time of return, especially of the patellar reflexes, marked differences are observable. The deep reflexes later are increased on the paralyzed side, and ankle clonus may be present. Plantar stimulation gives an extensor response in the great toe (Babinski's sign) or dorsal flexion of the foot on irritating the skin over the tibia (Oppenheim's sign). The other superficial reflexes are usually diminished. The sphincters are not affected.

The *course* depends upon the situation and extent of the lesion. If slight the hemiplegia may disappear completely within a few days or a few weeks. In severe cases the rule is that the leg gradually recovers before the arm, and the muscles of the shoulder girdle and upper arm before those of the forearm and hand. The face may recover quickly.

Except in the very slight lesions, in which the hemiplegia is transient changes take place which may be grouped as

**SECONDARY.**—These correspond to the chronic stage. In a patient in whom little or no improvement takes place within eight or ten weeks it will be found that the paralyzed limbs undergo certain changes. The leg, as a rule, recovers enough power to enable the patient to get about, although the foot is dragged. Occasionally a recurrence of severe symptoms is seen, even without a new hæmorrhage. In both arm and leg the condition of *secondary contraction* or *late rigidity* comes on, always most marked in the arm which becomes permanently flexed at the elbow and resists all attempts at extension. The wrist is flexed upon the forearm and the fingers upon the hand. The position of the arm and hand is very characteristic. There is frequently, as

the contractures develop, a great deal of pain. In the leg the contracture is rarely so extreme. The loss of power is most marked in the muscles of the foot and, to prevent the toes from dragging, the knee in walking is much flexed, or more commonly the foot is swung round in a half circle.

The reflexes are greatly increased at this stage. These contractures are permanent and incurable, and are associated with a secondary descending sclerosis of the motor path. There are instances, however, in which rigidity and contracture do not occur, but the arm remains flaccid, the leg having regained its power. This *hémiplegie flasque* of Bouchard is found most commonly in children. Among other secondary changes in late hemiplegia may be mentioned the following: Tremor of the affected limbs, post-paralytic chorea, the mobile spasm known as athetosis, arthropathies on the affected side, and muscular atrophy. The cool surface and thin glossy skin of a hemiplegic limb are familiar to all.

Atrophy of the muscles may occur, thought to be due in some cases to secondary alterations in the gray matter of the ventral horns; but atrophy may follow as a direct result of the cerebral lesion, the ventral horns remaining intact. In Quinke's case atrophy of the arm followed the development of a glioma in the anterior central convolution. The gray matter of the ventral horns was normal. These atrophies are most common in cortical lesions involving the domain of the third main branch of the Sylvian artery, and in central lesions involving the lenticulo-thalamic region. Their explanation is not clear. The wasting of cerebral origin, which occurs most frequently in children and leads to hemiatrophy of the muscles with stunted growth, is to be sharply separated from the hemiatrophy of the muscles of the adult following within a relatively short time upon the hemiplegia.

**Diagnosis.**—There are three groups of cases which offer difficulty:

(1) Cases in which the onset is gradual, a day or two elapsing before the paralysis is fully developed and consciousness completely lost, are readily recognized, though it may be difficult to determine whether the lesion is due to thrombosis or to hæmorrhage.

(2) In the sudden apoplectic stroke in which the patient rapidly loses consciousness the difficulty in diagnosis may be still greater, particularly if the patient is in deep coma when first seen.

The first point to be decided is the existence of hemiplegia. This may be difficult, although, as a rule, even in deep coma the limbs on the paralyzed side are more flaccid and drop instantly when lifted: whereas on the non-paralyzed side the muscles retain some degree of tonus. One cheek may puff or one side of the mouth splutter in expiration. The reflexes may be decreased or lost on the affected side and there may be conjugate deviation of the head and eyes. Rigidity in the limbs on one side is in favor of a hemiplegic lesion. It is practically impossible in a majority of these cases to say whether the lesion is due to hæmorrhage, embolism, or thrombosis.

(3) Large hæmorrhage into the ventricles or into the pons may produce sudden loss of consciousness with complete relaxation, simulating coma from uræmia, diabetes, alcoholism, opium poisoning, or epilepsy.

The previous history and the mode of onset may give valuable information. In *epilepsy* convulsions have preceded the coma; in *alcoholism* there is a history of constant drinking, while in *opium poisoning* the coma develops

more gradually, the pupils are small and the respirations slow. With *diabetic coma* the breath often smells of acetone. *Sunstroke*, pernicious forms of *malaria* and *illuminating gas poisoning* may give difficulty. In *ventricular hæmorrhage* the coma is sudden and comes on rapidly. The hemiplegic symptoms may be transient, quickly giving place to complete relaxation. Convulsions occur in many cases, and may be the very symptom to lead astray—as in a case of ventricular hæmorrhage in a puerperal patient, in whom, naturally enough, the condition was thought to be uræmic. Rigidity is often present. In hæmorrhage into the *pons* convulsions are frequent. The pupils may be strongly contracted, conjugate deviation may occur, and the temperature is apt to rise rapidly. The contraction of the pupils in pontine hæmorrhage naturally suggests opium poisoning. The difference in temperature in the two conditions is a valuable diagnostic point. The apoplecticiform seizures of general paresis have usually been preceded by abnormal mental symptoms, and the associated hemiplegia is seldom permanent.

The cerebral attacks in *Stokes-Adams disease* may resemble apoplexy very closely. One stout patient, the subject of many attacks, had been bled so often that he had a label inside his coat—"Do not bleed me in an attack."

It may be impossible at first to give a definite diagnosis. In emergency cases the physician should be particularly careful about the following points: The examination of the head for injury or fracture; the urine should be tested for albumin and sugar, and studied microscopically; a careful examination should be made of the limbs with reference to the degree of relaxation or the presence of rigidity, and the condition of the reflexes; the state of the pupils should be noted and the temperature taken. The odor of the breath (alcohol, acetone, chloroform, etc.) should be noted. The most serious mistakes are made in the case of patients who are drunk at the time of the attack, a combination by no means uncommon. Under these circumstances the case may erroneously be looked upon as one of alcoholic coma. It is best to regard each case as serious and to bear in mind that this is a condition in which, above all others, mistakes are common.

In *meningeal hæmorrhage*, as from ruptured aneurism, the attack is sudden, with pain in the head, rapid loss of consciousness, bilateral flaccidity or difficulty in determining the existence of hemiplegia, rapid rise in temperature, and the presence of blood under high pressure in the spinal fluid.

An apoplexy with a sudden onset with deep coma, increasing symptoms, fever and respiratory disturbance is usually due to hæmorrhage. A definite diagnostic point is the finding of blood in the cerebro-spinal fluid (care should be taken not to mistake blood due to the puncture as indicating hæmorrhage).

**Prognosis.**—From cortical hæmorrhage, unless very extensive, the recovery may be complete without a trace of contracture. This is more common when the hæmorrhage follows injury than when it results from disease of the arteries. Infantile meningeal hæmorrhage may produce idiocy or spastic diplegia. Large hæmorrhages into the corona radiata, and especially those which rupture into the ventricles, rapidly prove fatal. The hemiplegia which follows lesions of the internal capsule, the result of rupture of the lenticulo-striate artery, is usually persistent and followed by contracture. When the retro-lenticular fibres of the internal capsule are involved there may be hemi-

anæsthesia, and later, especially if the thalamus be implicated, hemichorea or athetosis.

In any case the following symptoms are of grave omen: deep coma at the onset; persistence or deepening of the coma during the second and third day; rapid rise in temperature within the first forty-eight hours after the initial fall. In the reaction which takes place on the second or third day the temperature usually rises, and its gradual fall on the third or fourth day with return of consciousness is a favorable indication. The rapid formation of bed-sores is a serious indication. The occurrence of albumin and sugar, if abundant, in the urine is an unfavorable symptom.

When consciousness returns and the patient is improving, the question is anxiously asked as to the paralysis. The extent of this can not be determined for some weeks. With slight lesions it may pass off entirely. If persistent at the end of a month some grade of permanent palsy is certain to remain, and gradually the late rigidity supervenes.

The *treatment* is discussed on page 1029.

## V. EMBOLISM AND THROMBOSIS

### (Cerebral Softening)

**Embolism.**—The embolus usually enters the carotid, rarely the vertebral artery. In the great majority of cases it comes from the left heart and is a vegetation of a fresh endocarditis or, more commonly, of a recurring endocarditis, or from the segments involved in an ulcerative process. Less often the embolus is a portion of a clot which has formed in the auricular appendix. Portions of clot from an aneurism, thrombi from atheroma of the aorta, or from the territory of the pulmonary veins, may also cause blocking of the branches of the circle of Willis. In the puerperal condition cerebral embolism is not infrequent. It may occur in women with heart disease, but in some instances the heart is uninvolved, and the condition is associated with the formation of clots in the heart or pulmonary veins. A majority of cases of embolism occur in heart disease, 89 per cent. (Saveliew). Cases are rare in the acute endocarditis of rheumatic fever, chorea, and febrile conditions. It is much more common in the secondary recurring endocarditis which attacks old sclerotic valves. The embolus most frequently passes to the left middle cerebral artery and the posterior cerebral and the vertebral are less often affected. A large plug may lodge at the bifurcation of the basilar. Embolism of the cerebellar vessels is rare.

Embolism occurs more frequently in women, owing, no doubt, to the greater frequency of mitral stenosis. Contrary to this general statement, in Newton Pitt's statistics of 79 cases at Guy's Hospital there were 44 males and 35 females. Saveliew gives 54 per cent. in women.

**Thrombosis.**—This occurs (1) about an embolus, (2) as the result of a lesion of the arterial wall (either endarteritis with or without atheroma or, particularly, the *syphilitic* arteritis), (3) in aneurisms, both large and miliary, and (4) as a direct result of abnormal conditions of the blood as in the anæmia of hæmorrhage, chlorosis, septicæmia and the cachexia of cancer. The arterial changes which lead to thrombosis and hæmorrhage are prac-

tically the same. Thrombosis occasionally follows ligation of the carotid artery. The thrombosis is most common in the middle cerebral and basilar arteries. It is suggested that softening of limited areas, sufficient to induce hemiplegia, may be caused by sudden collapse of cerebral arteries from cardiac weakness.

**Anatomical Changes.**—The embolus may retract, so that the blood flow is restored, or be carried on and lodge in a smaller vessel; in either case improvement occurs. In many instances a secondary thrombosis follows which may be extensive. In both embolism and thrombosis oedema occurs which, as in hæmorrhage, causes pressure and so is a cause of coma. Degeneration and softening of the territory supplied by the vessels are the ultimate result in both embolism and thrombosis. Blocking in a terminal artery may be followed by infarction, in which the territory may be deeply infiltrated with blood (hæmorrhagic infarction) or be pale, swollen, and necrotic (anæmic infarction). Gradually the process of *softening* proceeds, the tissue is infiltrated with serum and the nerve fibres degenerate and become fatty. The neuroglia is swollen and cedematous. The color of the softened area depends upon the amount of blood. The hæmoglobin undergoes gradual transformation, and the early red color may give place to yellow. Formerly much stress was laid upon the difference between *red*, *yellow*, and *white* softening. The red and yellow are seen chiefly on the cortex. Sometimes the red softening is particularly marked in cases of embolism and in the neighborhood of tumors. The gray matter shows many punctiform hæmorrhages. There is a variety of yellow softening—the *plaques jaunes*—common in elderly persons, occurring in the gray matter, in spots from 1 to 2 cm. in diameter, sometimes angular in shape, the edges cleanly cut, and the softened area represented by a turbid, yellow material. White softening occurs most frequently in the white matter, and is seen best about tumors and abscesses. Inflammatory changes are common in and about the softened areas. When the embolus comes from an infected focus, as in ulcerative endocarditis, suppuration may follow. The final changes vary greatly. The degenerated tissue elements are gradually removed, and if the region is small may be replaced by connective tissue and the formation of a scar. If large, the resorption results in the formation of a cyst.

The position and extent of the softening depend upon the obstructed artery. An embolus which blocks the middle cerebral at its origin involves not only the arteries to the anterior perforated space, but also the cortical branches, and in such a case there is softening in the neighborhood of the corpus striatum, as well as in part of the region supplied by the cortical vessels. The freedom of anastomosis between these branches varies. Thus, in embolism of the middle cerebral artery in which the softening has involved only the territory of the central branches, blood may reach the cortex through the anterior and posterior cerebrals. When the middle cerebral is blocked (as is perhaps oftenest the case) beyond the point of origin of the central arteries, one or other of its branches is usually most involved. The embolus may lodge in the vessel passing to the third frontal convolution, or in the artery of the ascending frontal or ascending parietal; or it may lodge in the branch passing to the supramarginal and angular gyri, or enter the lowest branch which is distributed to the upper convolutions of the temporal lobe. These are prac-

tically terminal arteries, and instances frequently occur of softening limited to a part, at any rate, of the territory supplied by them. Some of the most accurate focalizing lesions are produced in this way.

There is greater freedom of communication in the cortical branches of the different arteries than is usually admitted, but the absence of softening in some instances in which smaller branches are blocked shows how complete may be the compensation, probably by the capillaries. The dilatation of the collateral branches may take place very rapidly; thus a patient with chronic nephritis died twenty-four hours after the hemiplegic attack. There were recent vegetations on the mitral valve and an embolus in the right middle cerebral artery just beyond the first two branches. The central portion of the hemisphere was swollen and œdematous. The right anterior cerebral was greatly dilated, and its diameter was nearly three times that of the left.

**Symptoms.**—Extensive thrombotic softening may exist without any symptoms and it is not uncommon in post mortems on elderly persons to find areas of softening scattered over the convolutions. It may take place in the "silent" regions without exciting any symptoms. When the central or cortical branches of the middle cerebral arteries are involved the symptoms are similar to those of hæmorrhage from the same arteries. Permanent or transient hemiplegia results. When the central arteries are involved the softening in the internal capsule is commonly followed by hemiplegia. Certain peculiarities are associated with embolism and thrombosis respectively.

In *embolism* the patient is usually the subject of heart trouble, or there exist some of the conditions already mentioned. The onset is sudden, without premonitory symptoms but sometimes with intense headache. When the embolus blocks the left middle cerebral artery the hemiplegia is associated with aphasia. In *thrombosis*, on the other hand, the onset is more gradual; the patient has previously complained of headache, vertigo, tingling in the fingers; the speech may have been embarrassed for some days; the patient has had loss of memory or is incoherent, or paralysis begins at one part, as the hand, and extends slowly, and the hemiplegia may be incomplete or variable. Abrupt loss of consciousness is much less common, and when the lesion is small consciousness is retained. In thrombosis due to syphilitic disease, the hemiplegia may come on gradually without any disturbance of consciousness.

The hemiplegia following thrombosis or embolism has practically the characteristics, both primary and secondary, described under hæmorrhage.

The following may be the effects of blocking the different vessels: (a) *Vertebral*. The left branch is more frequently plugged. The effects are involvement of the nuclei in the medulla and symptoms of acute bulbar paralysis. It rarely occurs alone; more commonly with:

(b) *Blocking of the Basilar Artery*.—When this is entirely occluded, there may be bilateral paralysis from involvement of both motor paths. Bulbar symptoms may be present; rigidity or spasm may occur. The temperature may rise rapidly. The symptoms, in fact, are those of apoplexy of the pons.

(c) The *posterior cerebral* supplies the occipital lobe on its medial surface and the greater part of the temporo-sphenoidal lobe. If the main stem be thrombosed there is hemianopia with sensory aphasia. Localized areas of softening may exist without symptoms. Blocking of the main occipital branch (*arteria occipitalis* of Duret), or of the artery passing to the cuneus, may be

followed by hemianopia. Hemianæsthesia may result from involvement of the posterior part of the internal capsule. Symmetrical thrombosis of the occipital arteries of the two sides occurs, as in Förster's well-known case. Still more frequent is thrombosis of a branch of the posterior cerebral of one hemisphere and a branch of the middle cerebral of the other. In such cases the most pronounced instances of apraxia are met with.

(d) *Internal Carotid*.—The symptoms are variable. The vessel is ligated without risk in a majority of cases; in other instances transient hemiplegia follows; in others again the hemiplegia is permanent. These variations depend on the anastomoses in the circle of Willis. If these are large and free, no paralysis follows, but if the posterior and anterior communicating vessels are small or absent the paralysis may persist. In No. 7 of the Elwyn series of cases of infantile hemiplegia, the woman, aged twenty-four, when six years old, had the right carotid ligated for abscess following scarlet fever, with the result of permanent hemiplegia. Blocking of the internal carotid within the skull by thrombosis or embolism is followed by hemiplegia, coma, and usually death. The clot is rarely confined to the carotid itself, but spreads into its branches and may involve the ophthalmic artery.

(e) *Middle Cerebral*.—This is the vessel most commonly involved, and if plugged before the central arteries are given off, permanent hemiplegia usually follows from softening of the internal capsule. Blocking of the branches beyond this point may be followed by hemiplegia, which is more likely to be transient, involves chiefly the arm and face, and if the lesion be on the left side is associated with aphasia. There may be plugging of the individual branches passing to the inferior frontal (producing motor aphasia if the disease be on the left side), to the anterior and posterior central gyri (usually causing total hemiplegia), to the supramarginal and angular gyri (giving rise, if on the left side, probably without exception to the so-called visual aphasia (alexia), usually also to right-sided hemianopsia), or to the temporal gyri (in which event with left sided thrombosis word-deafness results).

(f) *Anterior Cerebral*.—No symptoms may follow, and even when the branches which supply the paracentral lobule and the top of the ascending convolutions are plugged the branches from the middle cerebral are usually able to effect a collateral circulation. Monoplegia of the leg may result. Hebetude and dullness of intellect may follow obstruction of the vessel.

**Prognosis.**—In embolism this depends on the extent of tissue which is deprived of blood supply; hence the collateral circulation is important. It is often impossible to decide this. In thrombosis the outlook is probably best in the syphilitic cases if early diagnosis and active treatment are possible. In cases due to other forms of arterial disease the ultimate outlook is grave as the lesions are likely to lead to further vascular damage. The outlook for the paralysis is much as in hæmorrhage.

**Treatment of Hæmorrhage, Thrombosis and Embolism.**—The chief difficulty is to determine whether the apoplexy is due to hæmorrhage or to thrombosis or embolism. The patient should be placed in bed, with his head moderately elevated and the neck free. He should be kept absolutely quiet. If there are dyspnoea, stertor, and signs of mechanical obstruction to respiration, he should be turned on his side. This lessens the liability to congestion of the lungs. Venesection seems to be indicated theoretically in cases of

hæmorrhage with high pressure, but practically is of little or no value and is not advisable as a rule. With marked cyanosis in plethoric subjects it is sometimes useful. As Cushing has shown experimentally, a rapid and increasing rise of arterial tension usually indicates an endeavor to counteract an increasing intracranial pressure. The indication under these circumstances is the relief of the intracranial pressure by craniotomy and removal of the clot, if this is possible. This is particularly applicable in subdural hæmorrhage. Horsley and Spencer, on experimental grounds, recommended the practice of compression of the carotid, particularly in the ingravescent form. An ice-bag may be placed on the head and hot bottles to the feet. The bowels should be freely opened, by calomel or elaterin followed by magnesium sulphate, which by mouth or rectum often gives marked relief to the symptoms of increased intracranial pressure from any cause. Counter-irritation to the neck or to the feet is not necessary. Catheterization may be necessary, especially if the patient remains long unconscious.

Special care should be taken to avoid bed-sores; and if bottles are used to the feet, they should not be too hot, since blisters may be readily caused by a much lower temperature than in health. Stimulants are not necessary, unless the pulse becomes feeble and signs of collapse supervene. During recovery the patient should be still kept entirely at rest, even in the mildest attacks remaining in bed for at least fourteen days. The ice-bag should still be kept to the head. The diet should be light and the bowels should be kept open. Attention should be paid to the position occupied by the paralyzed limb or limbs, which if swollen may be wrapped in cotton batting or flannel. Small doses of iodide (gr. v, 0.3 gm.) may be given.

In thrombosis or embolism venesection is not indicated, as it rather promotes clotting. If, as is often the case, the heart's action is feeble and irregular, small doses of digitalis may be given. The bowels should be kept open, but it is not well to purge actively, as in hæmorrhage.

In thrombosis with syphilitic disease of the arteries, most frequent in men between twenty and forty (in whom the hemiplegia often sets in without loss of consciousness), active antisymphilitic treatment is indicated; the iodide should be given in full dosage. Practically these are the only cases of hemiplegia in which we see satisfactory results from treatment.

Very little can be done for the *hemiplegia* which remains. The damage is too often irreparable and permanent, and it is very improbable that iodide of potassium, or any other remedy, hastens in the slightest degree Nature's dealing with the blood clot. Passive movements or massage, and later re-education should be used systematically, in order to maintain the nutrition of the muscles and to prevent contractures if possible. The massage should not be begun until at least ten days after the attack. The rubbing should be *toward* the body, and should not be continued for more than fifteen minutes at a time. It is doubtful if electricity is of much value; some regard it as more likely to do harm by increasing spasticity. The patient should be encouraged to perform simple movements and exercise himself and make attempts to walk when the acute features are over. When contractures occur, passive movements and massage are useful, and it has been suggested that tendon transplantation, or cross suture of nerves, may cause improvement. There is always the possibility of another attack and the patient should lead a quiet

even life with a simple diet, keeping the bowels open and avoiding undue exertion and emotional disturbance as much as possible.

In a case of complete hemiplegia the friends should at the outset be told frankly that the chances of full recovery are slight. Power is usually restored in the leg sufficient to enable the patient to get about, but in the majority of instances the finer movements of the hand are permanently lost. The general health should be looked after, the bowels regulated, and the secretions of the skin and kidneys kept active. In permanent hemiplegia in older persons some mental weakness often follows the attack, and the patient may become irritable and emotional. Lastly, when hemiplegia has persisted for more than three months and contractures have developed, it is the duty of the physician to explain to the patient or to his friends, that the condition is past relief, that medicines and electricity will do no good, and that there is no hope of cure.

#### VI. ANEURISM OF THE CEREBRAL ARTERIES

Miliary aneurisms are not included, but reference is made only to aneurism of the larger branches. The condition is not uncommon. There were 12 instances in 800 autopsies in the Montreal General Hospital. This is a considerably larger proportion than in Newton Pitt's collection from Guy's Hospital, 19 times in 9,000 inspections.

**Etiology.**—Males are more frequently affected than females. The disease is most common at the middle period of life. One of the Montreal cases was a lad of six and Pitt describes one at the same age. The chief causes are (*a*) congenital defects, (*b*) endarteritis, simple or syphilitic, which leads to weakness of the wall and dilatation, and (*c*) embolism. These aneurisms are often found with endocarditis. Pitt concludes that it is exceptional to find cerebral aneurism unassociated with fungating endocarditis. The embolus disappears and dilatation follows the secondary inflammatory changes in the coats of the vessel.

**Morbid Anatomy.**—The middle cerebral branches are most frequently involved. Except in one of 12 cases they were saccular and communicated with the lumen of the vessel by an orifice smaller than the circumference of the sac. In 166 cases (statistics of Osler, Lebert, Durand, and Bartholow) the middle cerebral was involved in 49, basilar in 44, internal carotid in 24, anterior cerebral in 14, posterior communicating in 8, anterior communicating in 11, vertebral in 7, posterior cerebral in 6, inferior cerebellar in 3. The size of the aneurism varies from that of a pea to that of a walnut. The hæmorrhage may be meningeal with very slight laceration of the brain substance, or, as Coats has shown, entirely within the substance.

**Symptoms.**—The aneurism may attain considerable size and cause no symptoms. In a majority of cases the first intimation is rupture and fatal apoplexy. Distinct symptoms are most frequently caused by aneurism of the internal carotid, which may compress the optic nerve or the commissure, causing neuritis or paralysis of the third nerve. A murmur may be audible. Aneurism in this situation may give rise to irritative and pressure symptoms at the base of the brain or bilateral temporal hemianopia. Aneurism of the vertebral or of the basilar may involve the nerves from the fifth to the twelfth.

A large sac at the termination of the basilar may compress the third nerves or the crura.

In some cases the aneurism ruptures through a very small opening and the blood escapes slowly. If this is into the meninges pain and stiffness of the neck may result. Intracranial pressure increases and signs of irritation follow with the picture of meningitis. The cerebro-spinal fluid is bloody and may contain altered pigment if the hæmorrhage has gone on for some days. The process tends to go on to death, but recovery has occurred.

The diagnosis is, as a rule, impossible. The larger sacs produce the symptoms of tumor, and their rupture is usually fatal. The treatment is that of cerebral hæmorrhage.

#### VII. THROMBOSIS OF THE CEREBRAL SINUSES AND VEINS

The condition may be primary or secondary. Lebert (1854) and Tonnele were among the first to recognize the condition clinically.

**Primary thrombosis** of the sinuses and veins is rare. It occurs (a) in children, particularly during the first six months of life, usually in connection with diarrhœa. Gowers believed that it is frequent, and that thrombosis of the veins is not an uncommon cause of infantile hemiplegia.

(b) With chlorosis and anæmia, the so-called *autochthonous sinus-thrombosis*. Of 82 cases of thrombosis in chlorosis, 78 were in the veins and 32 in the cerebral sinuses. The longitudinal sinus is most frequently involved. The thrombosis is usually associated with venous thromboses elsewhere and the patients die in one to three weeks, but recovery may occur.

(c) In the terminal stages of cancer, tuberculosis, and other chronic diseases thrombosis may occur in the sinuses and cortical veins. To the coagulum in these conditions the term *marantic thrombus* is applied.

**Secondary thrombosis** is much more frequent and follows extension of inflammation from contiguous parts to the sinus wall. The common causes are disease of the internal ear, fracture, compression of the sinuses by tumor, abscess or suppurative diseases outside the skull, particularly erysipelas, carbuncle, and parotitis. In secondary cases the lateral sinus is most frequently involved. Of 57 fatal cases in which ear disease caused death with cerebral lesions, there were 22 in which thrombosis existed in the lateral sinuses (Pitt). Tuberculous caries of the temporal bone may be responsible. The thrombus may be small, or fill the entire sinus and extend into the internal jugular vein. In more than half of these instances the thrombus was suppurating. The disease spreads directly from the necrosis on the posterior wall of the tympanum by the petroso-mastoid canal. It is not so common in mastoid disease.

**Symptoms.**—*Primary thrombosis* of the *longitudinal* sinus may occur without exciting symptoms and is found accidentally post mortem. There may be mental dulness with headache. Convulsions and vomiting may occur. In other instances there is nothing distinctive. In the chlorosis cases the head symptoms are usually marked. Ball's patient was dull and stupid, had vomiting, dilatation of the pupils, and double choked disks. Slight paresis of the left side occurred. An interesting feature in this case was the development of swelling of the left leg. In other cases the patients have headache,

vomiting, and delirium. Paralysis or paresis may be present. Bristowe reported a case in an anæmic girl of nineteen, who had convulsions, drowsiness, and vomiting. Tenderness and swelling developed in the position of the right internal jugular vein, and a few days later on the opposite side. The diagnosis was rendered definite by the occurrence of phlebitis in the right leg. The patient recovered. Such symptoms in anæmia should lead to the suspicion of cerebral thrombosis. In infants the diagnosis can rarely be made.

In thrombosis of the *cavernous* sinus there is prominence of the eyes with œdema of the orbit, conjunctiva and face. There is disturbance of vision with swelling of the disk and hæmorrhages. Paralysis of the ocular muscles may occur. With involvement of the *lateral* sinus the thrombus may extend to the jugular vein and be palpable.

In the *secondary thrombi* the symptoms are commonly those of septicæmia and in over 70 per cent. of Pitt's cases the mode of death was by pulmonary pyæmia. This author draws the following conclusions: (1) The disease spreads oftener from the posterior wall of the middle ear than from the mastoid cells. (2) The otorrhœa is generally of some standing, but not always. (3) The onset is sudden, the chief symptoms being pyrexia, rigors, pains in the occipital region and in the neck, associated with a septicæmic condition. (4) Well-marked optic neuritis may be present. (5) The appearance of acute local pulmonary mischief or of distant suppuration is almost conclusive of thrombosis. (6) The average duration is about three weeks, and death is generally from pulmonary pyæmia. The chief points in the diagnosis may be gathered from these statements.

Associated with thrombosis of the lateral sinus there may be venous stasis and painful œdema behind the ear and in the neck. The external jugular vein on the diseased side may be less distended than on the opposite side, since owing to the thrombus in the lateral sinus the internal jugular vein is less full than on the normal side, and the blood from the external jugular can flow more easily into it.

**Treatment.**—This from the medical side is symptomatic. Care should be taken not to allow clothing to press on the neck and to avoid bending the neck. Surgical measures should be carried out early. The secondary forms, especially those following upon disease of the middle ear, are often amenable to operation, and many lives have been saved by surgical intervention after extensive sinus thrombosis.

## VIII. CEREBRAL PALSIES OF CHILDREN

**Introduction.**—There are three great groups: I. Those due to *prenatal* factors, *agenesia cerebri*, *microcephalus*, *porencephaly*, *congenital cysts*, etc. II. *Natal* or *intrapartum* which includes the large group of birth palsies due to meningeal hæmorrhage, etc.; and III. The *post-natal* group of which the larger proportion is due to acute encephalitis occurring between the second and sixth year, and leading to hemiplegia. In all these cerebral palsies there are three important factors: (1) Disturbance in some degree of the normal mental development, (2) paralysis, and (3) spasticity in greater or less degree.

A number of important conditions may be grouped together for con-

venience of description—*aplasia cerebri*, meningeal hæmorrhage, spastic diplegia, Little's disease, bilateral athetosis, etc.

I. APLASIA (AGENESIA) CEREBRI.—This is due to failure of development of the cerebral cortex due to intra-uterine conditions. Nothing abnormal may be noted at birth, which has not been delayed or assisted by instruments. The head may be small and the sutures may close early. Then it is noticed that the child does not develop normally in the use of the muscles; the movements are irregular but not athetoid. The head wobbles, the child does not sit up, the dentition is delayed, and by the second year, the failure of development is evident. The arms and legs may become stiff and the condition of bilateral spastic rigidity supervene. More often the limbs remain relaxed, the child may learn to walk in an awkward way, the full power over the movements is never acquired, and the child settles into a state of idiocy. Anatomically the brain is small, the convolutions ill developed, and there may be areas of lobular sclerosis, sometimes the remarkable tuberoso form.

II. MENINGEAL HÆMORRHAGE (*With conservative paraplegia spastica cerebri* (Heine); *Little's disease*; *Tetraplegia spastica*).—Heine, one of the founders of modern orthopedics, recognized the cerebral origin of many of the palsies of children; and Little subsequently called attention to the "influence of abnormal parturition, difficult labors, premature birth and asphyxia on the mental and physical condition of the child, especially in relation to deformities." In 1885 Sarah McNutt's careful studies correlated the meningeal hæmorrhage with the subsequent palsies as recognized by Heine and Little.

The causes are: (1) Tearing of the veins from pressure on the head in a contracted pelvis and in forceps delivery. (2) Asphyxia. The extreme stasis, particularly just after the head is born, causes rupture of the veins at the point of entrance to the longitudinal sinus (Cushing). (3) Hæmorrhage may be in association with the hæmorrhagic condition of the newborn.

The hæmorrhage is from the pia, usually over the cortex and widely spread. It may be more on one side than the other, and may extend over the cerebellum. The brain substance may be softened or compressed, and present foci of hæmorrhage. The hæmorrhage may be extradural, and even extend into the spinal cord. First birth, premature birth, foot presentation, but above all, the indiscriminate and careless use of the forceps are the causal factors. There is much wisdom in the dread expressed by Shandy Senior, of the dangers of compression of the delicate and fine web of the brain.

**Symptoms.**—*Early.*—The asphyxia may be protracted. Unusual torpor, absence of the natural crying, inability to take the breast, flaccidity of the limbs, sometimes with rigidity on one side or convulsions, unequal and dilated pupils, and slow breathing with signs of atelectasis are the most suggestive features. There may be hæmorrhages elsewhere if the condition is associated with the hæmorrhagic disease of the newborn, as in cases reported by Green and by Margaret Warwick. Lumbar puncture may show blood.

*Late.*—If the child recovers, nothing may be noticed for a few months. Perhaps there are convulsions. The first thing to attract attention is that when the child should begin to walk the limbs are not used readily, and on examination a stiffness of the legs and arms is found. Even at the age of two the child may not be able to sit up, and often the head is not well supported by the neck muscles. The *rigidity*, as a rule, is more marked in the

legs, and there is an adductor spasm. When supported on the feet, the child either rests on its toes and the inner surface of the feet, with the knees close together, or the legs may be crossed. The stiffness of the upper limbs varies. It may be scarcely noticeable or the rigidity may be as marked as in the legs. When the spastic condition affects the arms as well as the legs, we speak of the condition as diplegia or tetraplegia; when the legs alone are involved, as paraplegia. There seems to be no sufficient reason for considering them separately. The spasticity is probably due to changes in the pyramidal system. Constant irregular movements of the arms are not uncommon. The child has great difficulty in grasping an object. The spasm and weakness may be more evident on one side than the other. Strabismus, nystagmus, unequal pupils and optic atrophy may occur. The mental condition is, as a rule, defective and convulsive seizures are common.

III. ACUTE SPORADIC ENCEPHALITIS OF CHILDREN WITH CONSECUTIVE HEMIPLEGIA.—This is an acute infection characterized by fever, convulsions, coma, and a consecutive hemiplegia.

**History.**—Heine first recognized *Hemiplegia spastica cereбрalis*, separating it from other forms of infantile paralysis.

**Etiology.**—Cases of hemiplegia in children's homes and institutions for the feeble-minded fall into two groups—(1) a large one, 95 out of 135 in Osler's series, in which the disease began about the second year, suddenly, in healthy children; and (2) a small one, with a more advanced age of onset, comprising cases of trauma; heart disease, etc. A certain number in the first group follow acute infections, especially whooping cough. The condition may follow a slight febrile illness during the first year. The incidence in relation to acute polio-myelitis is not known. There did not appear to be an increase of cases during the recent outbreaks.

**Pathology.**—The motor area of one hemisphere is involved in an acute hæmorrhagic lesion, the convolutions swollen and deeply injected, the veins thrombosed, and on section the substance is moist, deep red, and the limitation of the gray matter ill defined or obliterated. The picture corresponds with Strümpell's polio-encephalitis. When the patients come to autopsy years later, sclerosis with atrophy of the motor area is the most common lesion or there is a sub-meningeal cyst.

**Symptoms.**—Clinically the disease is sharply defined. A healthy child between the first and fifth years has a convulsion, or a series of them, with fever, possibly vomiting, and then becomes comatose. Preliminary indisposition is rare; headache may be complained of, but without warning the fit, as a rule, is the first symptom. The fever may reach 103°-104°. There may be marked conjugate deviation of the head and eyes: the pupils are usually dilated, and may be unequal. The head may be retracted, and naturally meningitis is suspected. In the deep coma the hemiplegia may be—often is—overlooked, but on careful examination the face is seen to be drawn and the arm and leg of one side limp and paralyzed. One of two things happens—the coma persists, the convulsions recur, and the child dies from the second to the fifth day, or the fever drops, the coma lessens, and within a few days the child seems well, but one side is paralyzed.

Complete recovery is rare. The face and arm improve rapidly, the legs lag and drag, as in an ordinary hemiplegia. Speech if disturbed returns.

The chief tragedy is a failure to develop mentally, which takes so many of these patients into the feeble-minded homes. The arm of the affected side may not develop but remains shorter and the hand smaller. In other cases recovery is not so complete; both leg and arm are spastic and the latter may present post-hemiplegic movements. Sensation is not disturbed. A distressing feature is the onset of *epilepsy*, which may be in the form of pure Jacksonian fits, *petit mal*, or general seizures. Of the 135 cases in the series, 41 had epilepsy.

*Post-hemiplegic Movements.*—It was in cases of this sort that Weir Mitchell first described the post-hemiplegic movements. They are extremely common, and were present in 34 of the series. There may be either slight tremor in the affected muscles, or inco-ordinate choreiform movements—the so-called post-hemiplegic chorea—or, lastly,

*Athetosis.*—This is a remarkable condition in which there is a combination of spasm with the most extraordinary bizarre movements of the muscles. The patient may not be able to walk. The head is turned from side to side; there are continual irregular movements of the face muscles, and the mouth is drawn and greatly distorted. The extremities are more or less rigid, particularly in extension. On the slightest attempt to move, often spontaneously, there are extraordinary movements of the arms and legs. The patients are often unable to help themselves on account of these movements. The reflexes are increased. The mental condition is variable.

IV. PARALYSIS DUE TO EMBOLISM.—This requires no special description as it is practically the same as in adults.

**Treatment.**—Cases with asphyxia and convulsions after difficult labors have been operated upon soon after birth by Cushing and others, and cortical clots have been removed. In some cases there has been a complete restoration to health and the usual spastic sequels have not occurred. If there is a hæmorrhagic tendency, the use of blood serum is advisable. As the child grows, conditions have to be met—the mental, requiring the care and training necessary for the grade of feeble-mindedness, and the orthopedic treatment of the spasticity, for which much can be done. Passive movements, massage and re-educational exercises are helpful. The educational care in institutions has shown how much patient training is able to help the development of these defective children. In all these patients the degree of development depends very much upon the thorough, painstaking and systematic training of their minds and muscles.

Surgically much may be done by tenotomy and the use of proper apparatus. For the relief of the spasticity operations on the brain are rarely of any help. Better results have been obtained by nerve resection, and where there is a high grade of bilateral spasticity, the resection of the posterior nerve roots appears sometimes to have been helpful.

The operation of “sympathetic ramisectomy” (Hunter and Royle), in which the gray rami communicantes to the nerve roots are divided or avulsed, results in marked decrease of spasticity in muscles in which there is an excess of “plastic tone” in the muscle fibres supplied by somatic nerves. The sympathetic supply to striated muscle does not reach the same muscle fibres as that from the lower motor neurones. The operation removes the efferent part of the reflex arc which maintains the plastic tonus. It does not decrease the

voluntary control but removes the sympathetic supply from the spastic muscles. It is most useful for the division of the sympathetic fibres to the limbs. The cases have to be carefully chosen and the best results should follow the operation in spasticity from cortical lesions.

#### IV. TUMORS, INFECTIONS, GRANULOMATA, AND CYSTS OF THE BRAIN

The following are the most common varieties of new growths within the cranium:

**Infectious Granulomata.**—(a) *Tubercle* may form large or small growths, usually multiple. Tuberculosis of the glands or bones may coexist, but the tuberculous disease of the brain may occur in the absence of other clinically recognizable tuberculous lesions. The disease is most frequent early in life. Three-fourths of the cases occur under twenty, and one-half of the patients are under ten years of age (Gowers). Of 300 cases of tumor in persons under nineteen collected from various sources by Starr, 152 were tubercle. The nodules are most numerous in the cerebellum and about the base.

(b) *Syphiloma* is most commonly found on the cortex cerebri or about the pons. The tumors are superficial, attached to the arteries or the meninges, and rarely grow to a large size. They may be multiple. A gummatous meningitis of the base is common and in this process the oculomotor nerves are often affected. The motor nerves of the eye are particularly prone to syphilitic infiltration, and ptosis and squint are common. The pituitary body may be involved with symptoms suggestive of diabetes insipidus.

**Tumors.**—(c) *Gliomas.*—These are the most common tumors and occur in all parts of the brain. They are often diffuse but may be encapsulated. The consistence may be much like that of the brain or they may be firm. They are usually very vascular and the vessels are liable to degeneration with resulting hæmorrhage, thrombosis and œdema. This often accounts for acute features appearing suddenly. The histological appearance varies considerably. They do not give rise to metastases in other organs. If necrosis occurs in the tumor, cyst formation may result.

(d) *Endothelioma* comes next in frequency and occurs most commonly in the membranes covering the hemispheres or brain stem, and for a long time may cause injury by its compression effects alone. When meningeal in origin, it is the form of tumor most amenable to surgical treatment. Tumors of this kind are particularly common in the cerebello-pontine recess.

(e) *Carcinoma* is almost always secondary to cancer in other parts. Cancerous tumors have been found in symmetrical parts of the brain.

(f) *Sarcoma.*—These are usually secondary if in the brain substance. Primary growths may arise from the meninges or cranium. They are usually encapsulated.

(g) Other varieties are fibroma, usually developing from the membranes; bony tumors, which grow sometimes from the falx, psammoma, cholesteatoma, neuroblastoma, neuroma, and angioma. Fatty tumors are occasionally found on the corpus callosum. There is a remarkable condition, originally described by Rokitsky, of brownish-black pigmentation of the brain, partly diffuse,

partly focal, associated with pigmented naevi of the skin. The naevi in the brain are in no sense a metastasis from the skin, but are benign tumors arising primarily (MacLachlan).

**Cysts.**—These occur between the membranes and the brain, as a result of hæmorrhage or softening. *Porencephalus* is a sequel of congenital atrophy or hæmorrhage, or may be due to a developmental defect. Hydatid cysts have been referred to in the section on parasites. An interesting variety of cyst is that which follows severe injury to the skull in early life. Gliomata often undergo cystic degeneration. Dermoid cyst has been described.

**Site.**—A majority of all tumors occur in the cerebrum and especially in the centrum ovale. The cerebellum, pons, and membranes are next most often involved. Glioma is more common in the hemispheres and grows slowly. It is usually single. Tubercles are usually multiple. Secondary sarcoma and carcinoma are often multiple.

**Symptoms and Signs.**—These vary greatly, depending on the position of the tumor, its size, rate of growth, and the occurrence of vascular changes, such as hæmorrhage, thrombosis and œdema. All tumors are without symptoms for a variable time; it is our business to recognize them at the earliest possible period. The symptoms are of two kinds. (1) *General*, due to increase in intracranial pressure, or (2) *local*, due to effects produced on parts of the nervous system. The following are the most important: *Headache*, either dull, aching, and continuous, or sharp, stabbing, and paroxysmal. It may be diffuse or limited to the back or front and is often remittent. When in the back of the head it may extend down the neck (especially in tumors in the posterior fossa), and when in the front it may be accompanied with neuralgic pains in the face. Occasionally the pain may be very localized and associated with tenderness on pressure. Pressure is the important factor in its production, and small or slowly growing tumors may not cause it.

*Choked disk* (optic neuritis, papilloedema) should be looked for in every patient presenting cerebral symptoms, for it is the first sign of increased intracranial pressure. Loss of visual acuity usually indicates that optic atrophy has set in. It is usually double, but occasionally is found in only one eye. Growths may attain considerable size without producing optic neuritis. On the other hand, it may occur with a very small tumor, when this tumor is so situated as to cause internal hydrocephalus. J. A. Martin, from an analysis of the literature with reference to the localizing value, concludes: When there is a difference in the amount of the neuritis in each eye it is more than twice as probable that the tumor is on the side of the most marked neuritis. It is constant in tumors of the corpora quadrigemina, present in 89 per cent. of cerebellar tumors, and absent in nearly two thirds of the cases of tumor of the pons, medulla, and of the corpus callosum. It is least frequent in cases of tuberculous tumor; most common in cases of glioma and cystic tumors.

Paton and Holmes report upon the eyes of 700 cases of cerebral tumor, concluding that the essential feature of the associated optic neuritis is œdema, and in 60 eyes examined histologically the one unfailing change was acute œdema, the origin of which they attribute to the venous engorgement.

*Vomiting* is a less constant feature but with headache and optic neuritis is of significance. Important points are the absence of definite relation to

meals and of digestive disturbances. It may be very obstinate, particularly in growths of the cerebellum and the pons.

*Giddiness* is often an early symptom, experienced on rising suddenly or turning quickly.

*Mental Symptoms.*—There is often some change, usually in the direction of dulness. Mania, depressive conditions, delusions, hallucinations and confusional states have been described but are usually late. The patient may act in an unnatural manner, or there may be stupor and heaviness. The patient may be emotional or silly, or there are symptoms resembling hysteria.

*Convulsions*, either general and resembling true epilepsy or localized (Jacksonian) in character. Seizures beginning with a gustatory or olfactory aura are common with tumors originating in the infundibular region.

*Circulation.*—Secondary to increased intracranial pressure there is increased blood pressure. The pulse rate is often slow. Respiration may be slow and sometimes is irregular.

**LOCAL FEATURES.**—The smaller the tumor and the less marked the general symptoms of cerebral compression the more likely is it that any focal symptoms occurring are of *direct* origin. Localizing features are often misleading. A frontal tumor may have cerebellar features due to increased intracranial pressure which has compressed the cerebellum against the base of the skull. The characteristic Bárány cerebellar tests have been present with temporal lobe tumors.

(a) *Central Motor Area.*—The symptoms are irritative or destructive in character. *Irritation* in the lower third may produce spasm in the muscles of the face, in the angle of the mouth, or in the tongue. The spasm with tingling may be strictly limited to one muscle group before extending to others, and this Seguin termed the *signal symptom*. The middle third of the motor area contains the centres controlling the arm, and here, too, the spasm may begin in the fingers, in the thumb, in the muscles of the wrist, or in the shoulder. In the upper third of the motor areas the irritation may produce spasm beginning in the toes, in the ankles, or in the muscles of the leg. In many instances the patient can determine accurately the point of origin of the spasm, and there are important sensory disturbances, such as numbness and tingling, which may be felt first at the region affected. It is important to determine, first, the point of origin, the *signal symptom*; second, the order or march of the spasm; and third, the subsequent condition of the parts first affected, whether it is a state of paresis or anæsthesia.

*Destructive* lesions in the motor zone cause paralysis, often preceded by local convulsive seizures; there may be a monoplegia, as of the leg, and convulsive seizures in the arm, often due to irritation. Tumors in the neighborhood of the motor area may cause localized spasms and subsequently, as the centres are invaded by the growth, paralysis occurs. With tumors in the left hemisphere the speech mechanism is apt to be involved if the transverse temporal gyrus or the third frontal convolution and their connecting path are implicated.

(b) *Prefrontal Region.*—Neither motor nor sensory disturbance may be present. The general symptoms are often well-marked. The most striking feature of growths in this region is a change in character and habits which may progress to stupor or dementia, particularly when the left side is in-

involved. In its extension downward the tumor may involve on the left side the lower frontal convolution and produce aphasia, or in its progress backward cause irritative or destructive lesions of the motor area. Exophthalmos on the side of the tumor may occur and be helpful in diagnosis.

(c) Tumors in the *parietal lobe*, particularly on the right side, may grow to a large size without causing any symptoms. There may be word-blindness and mind-blindness when the left angular gyrus and its underlying white matter are involved, and paraphasia. Astereognosis may accompany growths in the parietal lobe.

(d) Tumors of the *occipital lobe* produce hemianopia, and a bilateral lesion may produce blindness. Tumors in this region on the left hemisphere may be associated with word-blindness and mind-blindness. In all cases of tumor a careful study should be made of the fields of vision. In addition to the lateral hemianopia there may be remarkable visual hallucinations, and in tumors of the left occipital lobe dissociation of the color sense and inability to find the proper colors of various objects presented.

(e) Tumors in the *temporal lobe* may attain a large size without producing symptoms. In their growth they involve the lower motor centres. On the left side involvement of the transverse temporal gyri (auditory sense area) may be associated with word-deafness. Unpleasant hallucinations of taste and smell may occur.

(f) Tumors growing in the neighborhood of the *basal ganglia* produce hemiplegia from involvement of the internal capsule. Limited growths in either the nucleus caudatus or the nucleus lentiformis of the corpus striatum do not necessarily cause paralysis. Tumors in the thalamus opticus may also, when small, cause no symptoms, but, increasing, they may involve the fibres of the sensory portion of the internal capsule, producing hemianopia and sometimes hemianæsthesia. Growths in this situation are apt to cause early optic neuritis, and, growing into the third ventricle, may cause a distention of the lateral ventricles. What has been termed the *thalamic syndrome* may be present—hemianæsthesia to pain, touch and temperature, with the loss of deep sensibility. With this there may be a very remarkable type of pain, involving the hand and arm and the foot and leg, on the affected side, a sense of burning discomfort rather than sharp pain. Ataxic features are usually present and astereognosis. Motor hemiplegia may be present, and it is unaccompanied by contractures (Dana).

Growths in the *corpora quadrigemina* are rarely limited, but most commonly involve the crura cerebri as well. Ocular symptoms are marked. The pupil reflex is lost and there is nystagmus. In the gradual growth the third nerve is involved as it passes through the crus, in which case there will be oculo-motor paralysis on one side and hemiplegia on the other, a combination almost characteristic of unilateral disease of the crus.

(g) Tumors of the *pons* and *medulla*. The symptoms are chiefly those of pressure upon the nerves emerging in this region. In disease of the pons the nerves may be involved alone or with the pyramidal tract. Of 52 cases analyzed by Mary Putnam Jacobi, in 13 the cerebral nerves were involved alone, in 13 the limbs were affected, and in 26 there were hemiplegia and involvement of the nerves. In 22 of the latter there was alternate paralysis—i. e., involvement of the nerves on one side and of the limbs on the opposite

side. In 4 cases there were no motor symptoms. In tuberculosis (or syphilis) a growth at the inferior and inner aspects of the crus may cause paralysis of the third nerve on one side, and of the face, tongue, and limbs on the opposite side (syndrome of Weber). A tumor growing in the lower part of the pons usually involves the sixth nerve, producing internal strabismus, the seventh nerve, producing facial paralysis, and the auditory nerve, causing deafness. Conjugate deviation of the eyes to the side opposite that on which there is facial paralysis also occurs. When the motor cerebral nerves are involved the paralyzes are of the peripheral type (lower segment paralyzes).

Tumors of the *medulla* may involve the cerebral nerves alone or cause in some instances a combination of hemiplegia with paralysis of the nerves. Paralyzes of the nerves are helpful in topical diagnosis, but the fact must not be overlooked that one or more of the cerebral nerves may be paralyzed as a result of a much increased general intracranial pressure. Signs of irritation in the ninth, tenth, and eleventh nerves are usually present, and produce difficulty in swallowing, irregular action of the heart, irregular respiration, vomiting, and sometimes retraction of the head and neck. The hypoglossal nerve is least often affected. The gait may be unsteady or, if there is pressure on the cerebellum, ataxic. Occasionally there are sensory symptoms, numbness and tingling. Toward the end convulsions may occur.

(h) Tumors of the *cerebellum* usually give rise to very characteristic features, headache in the occipital region, giddiness, incoördination, nystagmus, ataxia on movement with difficulty in balancing and hence a staggering gait, weakness and hypotonia on the affected side, and early optic neuritis. The head may be tilted to the side of the tumor, and there may be a tendency to fall the same way. When the patient stands on one leg alternately the difference in steadiness is usually evident.

(i) Tumors of the cerebello-pontine angle show signs due to pressure on the cranial nerves with signs from the cerebellum when the tumor compresses it. The *acoustic* tumor, a neuro-fibroma of the eighth nerve, shows disturbance of the eighth with later involvement of the fifth and seventh nerves. Nystagmus occurs with signs of cerebellar involvement later.

Tumors or enlargements of the *pituitary gland* itself, or growths from a congenital *anlage* in its neighborhood which implicate the pituitary gland secondarily, are common. The congenital tumors arise presumably from developmental faults, and show either a teratomatous character or are solid or cystic tumors with squamous epithelium, often attaining adamantine characteristics. There are characteristic signs of pressure upon the neighborhood structures, bitemporal hemianopia being a frequent though not invariable feature. These lesions may occur in patients who have suffered from acromegaly, or who show signs of glandular deficiency or dyspituitarism, and in whom there may or may not be suggestive acromegalic tendencies. The X-rays are most useful in diagnosis.

**Diagnosis.**—The frequency of brain tumors should be remembered and a diagnosis made as early as possible. The occurrence of severe, perhaps intermittent, headache with choked disk is always suggestive. With definite general features and local signs the diagnosis should be fairly clear. But an early diagnosis is often very difficult. The patient should be carefully studied and a competent neurologist consulted if there is doubt. As pointed out by R. T.

**Williamson**, progressive hemiplegia, without other symptoms, a paralysis, which gradually becomes more marked day by day and week by week, is almost pathognomonic, even in the absence of optic neuritis, headache, and vomiting. Exceptions to this rule appear to be cerebral abscess, and rare instances of polio-encephalitis. It must not be forgotten that severe headache and choked disk may be caused by nephritis. The localization must be gathered from the consideration of the symptoms and from the data given in the section on Topical Diagnosis. Mistakes are most likely to occur in connection with uræmia, hysteria, vascular lesions, abscess, serous meningitis, hydrocephalus, and general paresis; but careful consideration of all the circumstances usually enables the practitioner to avoid error. The physicians of the National Hospital, London, have drawn attention to the importance of nasal irritation, as shown by the patients rubbing their noses frequently, as suggesting tumor. Care must be taken not to regard improvement under syphilitic treatment as positive evidence of syphilis, as this may occur with other tumors. Lumbar puncture should always be done with due consideration of the dangers and if necessary, as to exclude or prove syphilis, only a small amount of fluid should be removed. As to localization this may be very difficult if the growth is above the tentorium. In this the early signs are of more value than those of late stages. The X-ray study may be of value and should always be done. In about 45 of 100 cases of brain tumor the X-ray picture was of help in the diagnosis (Dandy). The introduction of air into the cerebral ventricles after the withdrawal of cerebro-spinal fluid may aid in the recognition of certain tumors by the X-ray study (pneumo-ventriculography).

**Prognosis.**—Syphilitic tumors alone are amenable to medical treatment. Tuberculous growths occasionally cease to grow and become calcified. The gliomata and fibromata, particularly when the latter grow from the membranes, may last for years. The general tendency is to progress; blindness results from optic neuritis and mental deterioration appears if the duration is sufficient. Death may be sudden, particularly in growths near the medulla; more commonly it is due to coma in consequence of increased intracranial pressure.

**Treatment.**—(a) MEDICAL.—Except in syphilis this is palliative only and in syphilis operation should be considered if the signs do not clear rapidly. A Wassermann test of the blood and cerebro-spinal fluid should always be made before antiluetic measures are instituted. Treatment with potassium iodide may cause a temporary amelioration of symptoms due to a glioma, so that the therapeutic test is not a dependable one. If syphilis is proved the iodide of potassium and mercury should be given. Arsphenamine is sometimes given in repeated small doses. The iodide should be given in increasing doses. In tuberculous tumors the outlook is less favorable, though instances of cure are reported, and there is post-mortem evidence that solitary tuberculous tumors may undergo changes and become obsolete. A general tonic treatment is indicated in these cases. The headache usually demands prompt treatment. Iodide of potassium in full doses sometimes gives marked relief and various analgesic drugs may be given. An ice-cap for the head or, in the occipital headache, the application of the Paquelin cautery may be tried. The bromides are not of much use in the headache from this cause, and, as the last

resort, morphia must be given. For the convulsions bromide of potassium is of little service.

(b) SURGICAL.—There is no doubt of the wisdom of referring these patients at once to men who make a special study of neurological surgery. Many tumors of the brain have been successfully removed. Though the percentage in which total enucleation is possible is doubtless small, yet marked amelioration of the pressure symptoms is possible by surgical measures. It is important that they should be instituted early. The most advantageous cases are the localized tumors growing from the dura and only compressing the brain substance. There have been numerous successful operations with removal of growths from the cerebellum and cerebello-pontine recess. For two objects the decompression operation may be performed, to relieve the headache, which it sometimes does promptly and permanently, and to save sight.

## V. INFLAMMATION OF THE BRAIN

### I. ACUTE ENCEPHALITIS

A focal or diffuse inflammation of the brain substance, usually of the gray matter (polio-encephalitis), is met with (a) as a result of trauma; (b) in certain intoxications, alcohol, food poisoning, and gas poisoning; (c) following the acute infections; (d) as one of the very rare varieties of polio-myelitis (page 972); and (e) epidemic encephalitis (page 974). The anatomical features are those of an acute hæmorrhagic polio-encephalitis, corresponding in histological details with acute polio-myelitis. Focal forms are seen in ulcerative endocarditis, in which the gray matter may present deep hæmorrhagic areas, firmer than the surrounding tissue. In the fevers there may be more extensive regions, involving two or three convolutions. This acute polio-encephalitis was thought by Strümpell to be the essential lesion in infantile hemiplegia. Localizing symptoms are usually present, though they may be obscured in the severity of the general infection. A most typical encephalitis accompanies the meningitis in cerebro-spinal fever.

In acute mania, delirium tremens, chorea insaniens, the maniacal form of exophthalmic goitre, and the so-called cerebral forms of fevers the gray cortex is congested, moist, and swollen, and with the finer methods of research will probably show changes which may be classed as encephalitis.

The *symptoms* are not very definite. In severe forms they are those of an acute infection; some cases have been mistaken for typhoid fever. The onset may be abrupt in an individual apparently healthy. Other cases have occurred in the convalescence from the fevers. The general symptoms are those which accompany all severe acute affections of the brain—headache, somnolence, coma, delirium, vomiting, etc. The *local* symptoms are very varied, depending on the extent of the lesions, and may be irritative or paralytic. Usually fatal within a few weeks, cases may drag on for weeks or months and recover, generally with paralysis. The *treatment* depends greatly on the primary condition.

*Traumatic* encephalitis, usually the result of a fall or blow on the head, shows hæmorrhages which may be on the surface or in the substance of the

brain, oedema and inflammatory reaction. The danger of increased intracranial pressure is always present. The general features may be marked and increase. Lumbar puncture often gives a blood-stained fluid. Decompression is indicated if the signs of increased intracranial pressure are definite.

## II. SUPPURATIVE ENCEPHALITIS: ABSCESS OF THE BRAIN

**Etiology.**—Suppuration of the brain substance is rarely primary, but results, as a rule, from extension of inflammation from neighboring parts or infection from a distance through the blood. Instances occur in which it is difficult to assign a cause. The largest number of cases occur between the twentieth and fortieth years, and the condition is more frequent in men than in women. In children under five years of age, the chief causes are otitis media and trauma. There are three important etiological factors.

(a) *Trauma.*—Falls upon the head or blows, with or without abrasion of the skin. More commonly it follows fracture or punctured wounds. In this group meningitis is frequently associated with the abscess. Simple trauma or concussion does not produce abscess but organisms may enter through a laceration of the base opening one of the many sinuses.

(b) By far the most important infective foci are those which arise in *direct extension from disease of the middle ear, of the mastoid cells, or of the accessory nasal sinuses.* From the roof of the mastoid antrum the infection readily passes to the sigmoid sinus and induces an infective thrombosis. In other instances the dura becomes involved, and a subdural abscess is formed, which may readily involve the arachnoid or the pia mater. In another group the inflammation extends along the lymph spaces, or the thrombosed veins, into the substance of the brain and causes suppuration. Macewen thinks that without local areas of meningitis the infective agents may be carried through the lymph and blood channels into the cerebral substance. Infection which extends from the roof of the tympanic cavity is most likely to be followed by abscess in the temporal lobe, while infection extending from the mastoid cells causes most frequently sinus thrombosis and cerebellar abscess.

(c) *In Septic Processes.*—Abscess of the brain is not often found in pyæmia. In ulcerative endocarditis multiple foci of suppuration are common. Localized bone disease and suppuration in the liver are occasional causes. Certain conditions in the lungs, particularly bronchiectasis, may be followed by abscess. It is an occasional complication of empyema. Abscess of the brain may follow the specific fevers.

**Morbid Anatomy.**—The abscess may be solitary or multiple, diffuse or circumscribed. Practically any one of the different varieties of pyogenic bacteria may be concerned. The bacteriological examination often shows different varieties. Occasionally cultures are sterile. In the acute, rapidly fatal cases following injury the suppuration is not limited; but in long standing cases the abscess is inclosed in a definite capsule, which may have a thickness of from 2 to 5 mm. The pus varies much in appearance, depending upon the age of the abscess. In early cases it may be mixed with reddish *débris* and softened brain matter, but in the solitary encapsulated abscess the pus is distinctive, having a greenish tint, an acid reaction, and a peculiar odor, sometimes like that of sulphuretted hydrogen. The brain substance surrounding

the abscess is usually oedematous and infiltrated. The size varies from that of a walnut to that of a large orange. There are cases in which the cavity occupies the greater portion of a hemisphere. Multiple abscesses are usually small. In four-fifths of all cases the abscess is solitary. Suppuration occurs most frequently in the cerebrum, and the temporal lobe is more often involved than other parts, and always on the side of the ear disease. The cerebellum is the next most common seat, particularly in connection with ear disease.

**Symptoms.**—These may be very obscure. Following injury or operation the disease may run an *acute* course, with fever, headache, delirium, vomiting, and rigors. The symptoms are those of suppurative meningo-encephalitis, and it may be very difficult to determine, unless there are localizing signs, whether there is really suppuration in the brain substance. In the cases following *ear disease* the symptoms may at first be those of meningeal irritation. There may be irritability, restlessness, severe headache, and aggravated earache. Other striking symptoms, particularly in the more prolonged cases, are drowsiness, slow cerebration, vomiting, and optic neuritis. In the chronic form which may follow injury, otorrhœa, or local lung trouble, there may be a latent period of weeks to several months, or even a year or more. In the "silent" regions, when the abscess becomes encapsulated there may be no symptoms whatever during the latent period. During this time the patient may be under careful observation and no suspicion of suppuration be aroused. Then severe headache, vomiting, and fever set in, perhaps with a chill. So, too, after a blow upon the head or a fracture the symptoms may be transient, and months afterward cerebral symptoms of the most aggravated character may develop.

The *localization* is often difficult. If situated in or near the motor region there may be convulsions or paralysis, and an abscess in the temporal lobe may compress the lower part of the pre-central convolution and produce paralysis of the arm and face, and on the left side cause aphasia. A large abscess may exist in the frontal lobe without causing paralysis, but in these cases there is almost always some mental dulness. In the temporal lobe, the common seat, there may be no focalizing symptoms. So also in the parieto-occipital region; though early examination may lead to the detection of hemianopia. In abscess of the cerebellum vomiting is common. If the middle lobe is affected there may be staggering—cerebellar inco-ordination. Localizing symptoms in the pons and other part are still more uncertain.

**Diagnosis.**—In the *acute* cases there is rarely any doubt. A consideration of possible etiological factors is of the highest importance. The history of injury followed by fever, marked cerebral symptoms, the onset of rigors, delirium, and perhaps paralysis, make the diagnosis certain. In chronic ear disease, such cerebral symptoms as drowsiness and torpor, with irregular fever, supervening upon the cessation of a discharge, should excite the suspicion of abscess. Cases in which suppurative processes exist in the orbit, nose, or naso-pharynx, or in which there has been subcutaneous phlegmon of the head or neck, parotitis, erysipelas, or tuberculous or syphilitic disease of the bones of the skull, should be carefully watched, and immediately investigated should cerebral symptoms appear. It is particularly in the *chronic* cases that difficulties arise. The symptoms resemble those of tumor of the brain; indeed, they are those of tumor plus fever. Choked disk, however, so commonly as

sociated with tumor, may be absent. In a patient with a history of trauma or with localized lung or pleural trouble, who for weeks or months has had slight headache or dizziness, the onset of fever, especially if intermittent and associated with rigors, intense headache, and vomiting, points strongly to abscess. The pulse rate in cerebral abscess is usually accelerated, but cases are not rare in which it is slowed. In all forms a leucocytosis may occur. Macewen lays stress upon the value of percussion of the skull as an aid in diagnosis. The note, which is uniformly dull, becomes much more resonant when the lateral ventricles are distended in cerebellar abscess and in conditions in which the venæ Galeni are compressed. Tenderness of the skull has been noted over the region of the abscess.

It is not always easy to determine whether the meninges are involved and often in ear disease the condition is a meningo-encephalitis. Sometimes with acute ear disease the symptoms simulate closely cerebral meningitis or abscess. Indeed, Gowers stated that not only may these general symptoms be produced by ear disease, but even distinct optic neuritis.

**Prognosis.**—This is always serious and the only hope is in successful drainage. Even then complications may appear and cause death.

**Treatment.**—In ear disease free discharge of the inflammatory products should be promoted and careful disinfection practised. The treatment of injuries and fractures comes within the scope of the surgeon. The acute symptoms, such as fever, headache, and delirium, must be treated by rest, an ice-cap, and sedatives. In all cases, when a reasonable suspicion exists of the occurrence of abscess, the brain should be explored. The cases following ear disease, in which the suppuration is in the temporal lobe or in the cerebellum, offer the most favorable chances. The localization can rarely be made accurately and the operator must be guided more by general anatomical and pathological knowledge. In cases of injury the exploration should be over the seat of the blow or the fracture. Hexamine should be given in full dosage.

## VI. HYDROCEPHALUS

**Definition.**—A condition, congenital or acquired, in which there is a great accumulation of fluid within the ventricles of the brain. The cases may be divided into three groups—idiopathic internal hydrocephalus (serous meningitis), congenital or infantile, and secondary or acquired.

**Serous Meningitis** (*Idiopathic Internal Hydrocephalus; Angio-neurotic Hydrocephalus*).—A knowledge of this condition explains many anomalous and puzzling cases. An ependymitis causing a serous effusion into the ventricles, with distention and pressure effects, it may be compared to the serous exudates in the pleura or synovial membranes. It is not certain that the process is inflammatory, and Quinke likens it to angio-neurotic oedema. In very acute cases the ependyma may be smooth and natural looking; in more chronic cases thickened and sodden.

Both children and adults are affected, the latter more frequently. In the acute form the condition is mistaken for tuberculous or purulent meningitis. There are headache, retraction of the neck, and signs of increased intracranial pressure, choked disk, slow pulse, etc. Fever is usually absent, but there are

cases with recurring paroxysms. In the chronic form the features are those of tumor—general, such as headache, slight fever, somnolence and delirium; and local, as exophthalmos, optic neuritis, spasms and rigidity of muscles, and paralysis of the cerebral nerves. Exacerbations occur and the symptoms vary in intensity. Recovery may follow and some reported cases of disappearance of symptoms of brain tumor belong in this category.

□ A variety of this is the *circumscribed serous meningitis* confined to the cerebello-pontile angle, due to adhesions of the arachnoid to the cerebellum in the region of the flocculus. Fluid accumulates in the cisterna lateralis, which has its own choroid plexus. The increased pressure leads to disturbance in function of the nerves in this region, causing the syndrome described by Bárány—tinnitus with deafness, vertigo, occipital headache, facial paralysis, and the “pointing error.” Other lesions of this region, syphilitic meningitis and tumors, may, of course, cause this syndrome.

**Chronic Internal Hydrocephalus.**—This may be due to obstruction of outflow from the ventricles in the aqueduct of Sylvius (a congenital anomaly) or in the foramina of Magendie and Luschka (usually from inflammation) or to decreased absorption from the sub-arachnoid space (Dandy and Blackfan).

The lateral ventricles are enormously distended, but the ependyma is usually clear, sometimes a little thickened and granular, and the veins large. The choroid plexuses are vascular, sometimes sclerotic, but often natural looking. The third ventricle is enlarged, the aqueduct of Sylvius dilated, and the fourth ventricle may be distended. The quantity of fluid may reach several litres. It is limpid and contains a trace of albumin and salts. The changes in consequence of the ventricular distention are remarkable. The cerebral cortex is greatly stretched, and over the middle region the thickness may amount to no more than a few millimetres without a trace of the sulci or convolutions. The basal ganglia are flattened. The skull enlarges, and the circumference of the head of a child of three or four years may reach 25 or even 30 inches. The sutures widen. Wormian bones develop in them, and the bones of the cranium become exceedingly thin. The veins are marked beneath the skin. A fluctuation wave may sometimes be obtained, and Fisher's brain murmur may be heard. The orbital plates of the frontal bone are depressed, causing exophthalmos, so that the eyeballs can not be covered by the eyelids. The small size of the face, widening somewhat above, is striking in comparison with the enormously expanded skull.

The enlarged head may obstruct labor; more frequently the condition is noticed some time after birth. It has occurred in several members of the same family. Convulsions may occur. The reflexes are increased, the child learns to walk late, and ultimately in severe cases the legs become feeble and sometimes spastic. Sensation is much less affected than motility. Choked disk is not uncommon. The mental condition is variable; the child may be bright, but, as a rule, there is some grade of imbecility. The congenital cases usually die within the first four or five years. The process may be arrested and the patient may reach adult life. Even when extreme, the mental faculties may be retained, as in Bright's celebrated patient, Cardinal, who lived to the age of twenty-nine, and whose head was translucent when the sun was shining behind him. Care must be taken not to mistake the rachitic head for hydro-

cephalus. The condition may be associated with other defects, harelip, spina bifida and club-foot.

Dandy has introduced a method of fluoroscopy after the injection of air into the ventricles, the outlines of which are then well seen and the extent but not always the type of hydrocephalus determined.

**Acquired Chronic Hydrocephalus.**—This is stated to be occasionally primary (idiopathic)—that is to say, it comes on spontaneously in the adult without observable lesion. Dean Swift is said to have died of hydrocephalus, but this seems unlikely. It is based upon the statement that "he (Mr. White-way) opened the skull and found much water in the brain," a condition no doubt of *hydrocephalus ex vacuo*, due to the wasting associated with his prolonged illness and paralysis. In nearly all cases there is either a tumor at the base of the brain or in the third ventricle, which compresses the venæ Galeni. The passage from the third to the fourth ventricle may be closed, either by a tumor or by parasites. The foramen of Magendie may become closed by meningitis. Chronic inflammation of the ependyma may block the foramina of exit of the ventricular fluid. There may be unilateral hydrocephalus from closure of one of the foramina of Monro. In cerebro-spinal fever the foramina of exit of the fluid may be occluded, with great distention of the ventricles. These conditions in adults may produce the most extreme hydrocephalus without any enlargement of the head. Even when the tumor begins early in life there may be no expansion of the skull. In a girl aged sixteen, blind from her third year, the head was not unusually large, the ventricles were enormously distended, and in the Rolandic region the brain substance was only 5 mm. in thickness. A tumor occupied the third ventricle. In other instances the sutures separate and the head gradually enlarges.

The *symptoms* are curiously variable. In the case mentioned there were headaches and gradual blindness; then a prolonged period in which she was able to attend to her studies. Headaches again supervened, the gait became irregular and somewhat ataxic. Death occurred suddenly. In another case there were prolonged attacks of coma with a slow pulse, and on one occasion the patient remained unconscious for more than three months. Gradually progressing optic neuritis without focalizing symptoms, headache, and attacks of somnolence or coma are suggestive symptoms. These cases of acquired chronic hydrocephalus can not be certainly diagnosed during life, though the condition may be suspected. They simulate tumor very closely.

**Diagnosis.**—There is no difficulty if enlargement of the head is present. The forms secondary to meningitis are usually easily recognized. If due to growths in the brain stem the signs are generally marked. In adults it is difficult to distinguish from intracranial growth.

**Prognosis.**—This depends largely on the cause. If the course is acute and progressive the outlook is hopeless. In mild cases recovery may follow but there is a danger of mental defect or epilepsy. Remissions of the symptoms may occur, especially in cases of obscure origin.

**Treatment.**—The use of mercury and iodides is advised by some writers. Various operations have been devised for conveying the fluid to the subtemporal or subcutaneous regions, or even connecting the cisterna magna directly with the longitudinal sinus.

Decompression may be advisable in some cases.

## J. DISEASES OF THE PERIPHERAL NERVES

## I. NEURITIS

Neuritis may be *localized* in a single nerve, or *general*, involving a large number of nerves—*multiple neuritis* or *polyneuritis*.

**Etiology.**—*Local neuritis* arises from (a) *cold*, which is a frequent cause, as, for example, in the facial nerve. (b) *Trauma*—wounds, blows, direct pressure on the nerves, the tearing and stretching which follow a dislocation or a fracture, and the hypodermic injection of ether. Under this come the professional palsies, due to pressure in the exercise of certain occupations. (c) *Extension of inflammation* from neighboring parts, as in a neuritis of the facial nerve due to caries in the temporal bone or in syphilitic disease of the bones, arthritis, and occasionally in tumors.

*Multiple neuritis* has a very complex etiology, the causes of which may be classified as follows: (a) The toxins of *infectious* diseases, as in leprosy, diphtheria, typhoid fever, small-pox, etc.; (b) the *organic poisons*, such as alcohol and ether, bisulphide of carbon and naphtha, and metallic bodies, as lead and arsenic; (c) *cachectic* conditions, such as occur in anæmia, cancer, tuberculosis, or marasmus from any cause; (d) the *endemic* neuritis or beriberi; and (e) lastly, there are cases in which none of these factors prevail, but the disease sets in suddenly after overexertion or exposure to cold.

**Morbid Anatomy.**—In neuritis due to the extension of inflammation the nerve is usually swollen, infiltrated, and red in color. The inflammation may be chiefly *perineural* or it may pass into the deeper portion—*interstitial* neuritis—in which form there is an accumulation of lymphoid elements between the nerve bundles. The nerve fibres themselves may not appear involved, but there is an increase in the nuclei of the sheath of Schwann. The myelin is fragmented, the nuclei of the internodal cells are swollen, and the axis-cylinders present varicosities or undergo granular degeneration. Ultimately the nerve fibres may be completely destroyed and replaced by a fibrous connective tissue in which much fat is sometimes deposited—*lipomatous neuritis*.

The other form is *parenchymatous* neuritis, in which the changes are like those in the secondary or Wallerian degeneration, which follows when the nerve fibre is cut off from the cell body of the neurone to which it belongs. The medullary substance and the axis-cylinders are chiefly involved, the interstitial tissues being but little altered or only affected secondarily. The muscles connected with the degenerated nerves usually show marked atrophic changes, and in some instances the change in the nerve sheath appears to extend directly to the interstitial tissue of the muscles.

**Symptoms.**—**LOCALIZED NEURITIS.**—As a rule, the constitutional disturbances are slight. The most important symptom is *pain* of a boring or stabbing character, usually felt in the course of the nerve and in the parts to which it is distributed. The nerve itself is sensitive to pressure, probably, as Weir Mitchell suggested, owing to the irritation of its *nervi nervorum*. The skin may be slightly reddened or even œdematous over the seat of the inflammation. Mitchell described increase in the temperature and sweating in the affected

region, and such disturbances as effusion into the joints and herpes. The function of the muscle to which the nerve fibres are distributed is impaired, motion is painful, and there may be twitchings or contractions. The *sensation* of the part may be somewhat deadened, even when the pain is greatly increased. In the more chronic cases of local neuritis, such, for instance, as follow the dislocation of the humerus, the localized pain, which at first, may be severe, gradually disappears, though some sensitiveness of the brachial plexus may persist for a long time, and the nerve cords may be firm and swollen. The *pain* is variable—sometimes intense and distressing; in others not causing much inconvenience. Numbness and formication may be present and tactile sensation greatly impaired. The motor disturbances are marked. Ultimately there is extreme atrophy of the muscles. Contractures may occur in the fingers. The skin may be reddened or glossy, the subcutaneous tissue œdematous, and the nutrition of the nails may be defective. In some cases subcutaneous fibroid nodules may develop.

A neuritis limited at first to a peripheral nerve may extend upward—the so-called ascending or migratory neuritis—and involve the larger nerve trunks, or even reach the spinal cord—causing subacute myelitis (Gowers). The condition is rarely seen in the neuritis from cold, or in that which follows fevers; but it occurs most frequently in traumatic neuritis.

J. K. Mitchell, in his monograph on injuries of nerves, concluded that the larger nerve trunks are most susceptible, and that the neuritis may spread either up or down, the former being the most common. The paralysis secondary to visceral disease, as of the bladder, may be due to an ascending neuritis. The inflammation may extend to the nerves of the other side, either through the spinal cord or its membranes, or without any involvement of the nerve-centres, the so-called sympathetic neuritis. The *electrical* changes in localized neuritis vary a great deal, depending upon the extent to which the nerve is injured. The lesion may be so slight that the nerve and the muscles to which it is distributed may react normally to both currents; or it may be so severe that the typical reaction of degeneration develops within a few days—i. e., the nerve does not respond to stimulation by either current, while the muscle reacts only to the galvanic current and in a peculiar manner. The contraction caused is slow and lazy, instead of sharp and quick as in the normal muscle, and the AC contraction is usually stronger than the KC contraction. Between these extremes there are many grades, and a careful electrical examination is an important aid to diagnosis and prognosis.

The duration varies from a few days to weeks or months. A slight traumatic neuritis may pass off in a day or two, while the severer cases, such as follow unreduced dislocation of the humerus, may persist for months or never be completely relieved.

**MULTIPLE NEURITIS.**—The following are the most important groups:

(a) *Acute Febrile Polyneuritis.*—The attack follows exposure to cold or overexertion, or, in some instances, comes on spontaneously. The onset resembles that of an acute infectious disease. There may be a definite chill, pains in the back and limbs or joints, so that the case may be thought to be rheumatic fever. The temperature rises rapidly and may reach 103° or 104° F. There are headache, loss of appetite, and the general symptoms of acute infection. The limbs and back ache. Intense pain in the nerves, how-

ever, is by no means constant. Tingling and formication are felt in the fingers and toes, and there is increased sensitiveness of the nerve trunks or of the entire limb. Loss of muscular power, first marked, perhaps, in the legs, gradually comes on and extends with the features of an ascending paralysis. In other cases the paralysis begins in the arms. The extensors of the wrists and the flexors of the ankles are early affected, so that there is foot and wrist drop. In severe cases there is general loss of muscular power, producing a flabby paralysis, which may extend to the muscles of the face and to the intercostals, and respiration may be carried on by the diaphragm alone. The muscles soften and waste rapidly. There may be only hyperæsthesia with soreness and stiffness of the limbs; in some cases, increased sensitiveness with anæsthesia; in other instances the sensory disturbances are slight. The Argyll-Robertson pupil may be present and the pupils may be unequal. Involvement of the cranial nerves is rare, but the third, fifth and seventh have been involved. The vagus may be attacked and the quickening of the pulse is usually attributed to this cause. Involvement of the bladder and rectum is rare, but it does occur and does not necessarily mean involvement of the cord. The clinical picture is not to be distinguished, in many cases, from Landry's paralysis; in others, from the subacute myelitis of Duchenne.

The *course* is variable. In the most intense forms the patient may die in a week or ten days, with involvement of the respiratory muscles or from paralysis of the heart. As a rule, in cases of moderate severity, after persisting for five or six weeks, the condition remains stationary and then slow improvement begins. The paralysis in some muscles may persist for many months and contractures may occur from shortening of the muscles, but even when this occurs the outlook is, as a rule, good, although the paralysis may have lasted for a year or more.

(b) *Recurring Multiple Neuritis*.—Under the term *polyneuritis recurrens* Mary Sherwood described from Eichhorst's clinic 2 cases in adults—in one case involving the nerves of the right arm, in the other both legs. In one patient there were three attacks, in the other two, the distribution in the various attacks being identical.

(c) *Alcoholic Neuritis*.—This, perhaps the most important form of multiple neuritis, was graphically described in 1822 by James Jackson, Sr., of Boston. Wilks recognized it as alcoholic paraplegia, but the starting point of the recent researches dates from the observations of Dumesnil, of Rouen. It occurs most frequently in women, particularly in steady, quiet tipplers. Its appearance may be the first revelation of habits of secret drinking. The onset is usually gradual, and may be preceded for weeks or months by neuralgic pains and tingling in the feet and hands. Convulsions are not uncommon. Fever is rare. The *paralysis* gradually sets in, at first in the feet and legs, and then in the hands and forearms. The extensors are affected more than the flexors, so that there is wrist-drop and foot-drop. The paralysis may be thus limited and not extend higher in the limbs. In other instances there is paraplegia alone, while in some extreme cases all the extremities are involved. In rare instances the facial muscles and the sphincters are affected. The *sensory* symptoms are very variable. There are cases in which there are numbness and tingling only, without great pain. In other cases there are severe burning or boring pains, the nerve trunks are sensitive, and the mus-

cles are sore when grasped. The hands and feet are frequently swollen and congested, particularly when held down for a few moments. The cutaneous reflexes, as a rule, are preserved. The deep reflexes are usually lost.

The *course* of these alcoholic cases is, as a rule, favorable, and after persisting for weeks or months improvement gradually begins, the muscles regain their power, and even in the most desperate cases recovery may follow. The extensors of the feet may remain paralyzed for some time, and give to the patient a distinctive walk, the so-called *steppage* gait, characteristic of peripheral neuritis. It is sometimes known as the pseudo-tabetic gait, although it could not well be mistaken for the gait of ataxia. The foot is thrown forcibly forward, the toe lifted high in the air so as not to trip upon it. The entire foot is slapped upon the ground as a flail. It is an awkward, clumsy gait, and gives the patient the appearance of constantly stepping over obstacles. Among the most striking features are the *mental* symptoms. Delirium is common, and there may be hallucinations with extravagant ideas, somewhat like those of general paresis. In some cases the picture is that of delirium tremens, but the most peculiar and almost characteristic mental disorder is that so well described by Wilks, in which the patient loses all appreciation of time and place, and describes with circumstantial details long journeys which, he says, he has recently taken, or tells of persons whom he has just seen. This is the so-called Korsakoff's syndrome or psychosis.

(d) *Multiple Neuritis in the Infectious Diseases.*—This has been already referred to, particularly in diphtheria, in which it is most common. The peripheral nature of the lesion in diphtheria has been shown by post mortem examination. The outlook is usually favorable and, except in diphtheria, fatal cases are uncommon. Multiple neuritis in tuberculosis, diabetes, and syphilis is of the same nature, being probably due to toxæmia of some kind. It may follow suppuration anywhere, as septic sore throat, and in the recent war multiple neuritis was seen not infrequently after superficial septic sores.

(e) *The Metallic Poisons.*—Neuritis from *arsenic* may follow: (1) The medicinal use particularly of Fowler's solution. In one case of Hodgkin's disease general neuritis was caused by 10 drams of the solution. In chorea a good many cases have been reported. Changes in the nails are not uncommon, chiefly the transverse ridging. In the case of a young woman who had taken "rough-on-rats" there were remarkable white lines—the leuconychia—running across the nails, without any special ridging. C. J. Aldrich finds that this is not uncommon in chronic arsenical poisoning. (2) The accidental contamination of food or drink. Chrome yellow may be used to color cakes, as in the cases recorded by D. D. Stewart. A remarkable epidemic of neuritis occurred in the Midland Counties of England, which was traced to the use of beer containing small quantities of arsenic, a contamination from the sulphuric acid used in making glucose. (3) A single dose taken by accident or purposely. Pigmentation of the skin is an important distinguishing sign. *Lead* is a much more frequent cause. Neuritis has followed the use of mercurial inunctions. Zinc is a rare cause. In a case seen with Urban Smith neuritis followed the use of two grains of the sulpho-carbolate taken daily for three years. Tea, coffee, and tobacco are mentioned as rare causes.

(f) *Endemic neuritis, beri-beri*, is considered elsewhere.

**ANÆSTHESIA PARALYSIS.**—Here may most appropriately be considered the forms of paralysis following the use of anæsthetics, or of too long-continued compression during operations. There are two groups of cases:

(a) During an operation the nerves may be compressed, either the brachial plexus by the humerus or the musculo-spiral by the table. The pressure most frequently occurs when the arm is elevated alongside the head, as in laparotomy done in the Trendelenburg position, or held out from the body, as in breast amputations. Instances of paralysis of the crural nerves by leg-holders are also reported. The too firm application of a tourniquet may be followed by a severe paralysis.

(b) Paralysis from cerebral lesions during etherization. Apoplexy or embolism may occur during anæsthesia. In Montreal a cataract operation was performed on an old man. He did not recover from the anæsthetic and post mortem a cerebral hæmorrhage was found. Epileptic convulsions may occur during anæsthesia, and prove fatal. The possibility of paralysis from loss of blood in operations has to be considered. Paralysis might result from the toxic effects of ether in a very protracted administration.

**ANGIOPATHIC PARALYSIS.**—Digital compression, the protracted application of the tourniquet and ligation of the main vessel taught us that normal action of the nerves and muscles of a limb was dependent upon a good blood supply. In sudden blocking of the femoral artery with an embolus, the pain is not simply in the site of the blockage, but is more or less diffuse throughout the limb, which the patient moves very slightly and with the greatest difficulty. In the recent war in the numerous injuries to the arteries, these angiopathic paralysees were not uncommon. In a study of ten cases of severe wounds of the main vessel of a limb Burrows found the chief symptoms to be: (a) subjective changes, numbness, tingling, etc.; (b) anæsthesia, usually of the glove type; (c) paralysis, often complete; (d) hardness of the muscles, and (e) œdema of the limb. With the re-establishment of the collateral circulation these may disappear in a few days.

**Diagnosis.**—The diagnosis of neuritis is often made carelessly and from nothing more than the complaint of pain. Pain in neuritis occurs only when a sensory or mixed nerve is involved. There is pain in the course of the nerve with tenderness or pain on pressure on it, changes in sensation in the area supplied, alteration in the deep reflexes, and muscular weakness and atrophy with altered electrical reactions. A loss of faradic irritability and a marked decrease in galvanic irritability are important signs of multiple neuritis. The relation of the motor and sensory changes is variable. Among the conditions which may lead to error are diseases of the cord, meninges or posterior nerve roots, spondylitis, arthritis, fibrositis, anatomical anomalies, wrong posture, perhaps with muscular strain, and flatfoot.

There is rarely any difficulty in distinguishing the alcohol cases. The combination of wrist- and foot-drop with congestion of the hands and feet, and the peculiar delirium are quite characteristic. The rapidly advancing cases with paralysis of all extremities, often reaching to the face and involving the sphincters, are more commonly regarded as of spinal origin, but the general opinion seems to point strongly to the fact that all such cases are peripheral. The less acute cases, in which the paralysis gradually involves the legs and arms with rapid wasting, simulate closely and are usually con-

founded with the subacute atrophic spinal paralysis of Duchenne. The diagnosis from tabes is rarely difficult. The *steppage* gait is entirely different. There is rarely positive incoördination and the station is good. Foot-drop is not common in tabes. The lightning pains are absent and there are usually no pupillary symptoms. The study of the spinal fluid is usually definite. The etiology is of moment.

**Treatment.**—Rest in bed is essential and the further ingestion of a causal agent should be prevented. In the acute cases with fever the salicylates and antipyrin are recommended. To allay the intense pain morphia or hot applications of lead water and laudanum are often required. Great care must be exercised in treating the alcoholic form, and the physician must not allow himself to be deceived by the statements of the relatives. It is sometimes exceedingly difficult to get a history of drinking. If there is any tendency to bed-sores an air-bed should be used or the patient placed in a continuous bath. Gentle friction of the muscles may be applied from the outset, and in the later stages, when the atrophy is marked and the pains have lessened, massage is probably the most reliable means at our command. Contractures should be prevented or if present gradually overcome by passive movements and extension. Often with the most extreme deformity from contracture, recovery is, in time, still possible. The interrupted current is useful when the acute stage is passed. Of internal remedies, strychnia is of value and may be given in increasing doses. If there is a suspicion of syphilis the usual treatment should be given.

## II. NEUROMATA

Tumors situated on nerve fibres may consist of nerve substance proper, the true neuromata, or of fibrous tissue, the false neuromata. The true neuroma usually contains nerve fibres only, or in rare instances ganglion cells. Cases of ganglionic or medullary neuroma are rare; some of them, as Lancereaux suggests, are undoubtedly instances of malformation of the brain substance. In other instances the tumor is, probably, a glioma with cells closely resembling those of the central nervous system. The growths are often intermediate in anatomical structure between the true and the false.

**Plexiform Neuroma.**—In this remarkable condition the various nerve cords may be occupied by many hundreds of tumors. The cases are often hereditary and usually congenital. The tumors may occur in all the nerves of the body, and, as numbers of them may be made out on palpation, the diagnosis is usually easy. One of the most remarkable cases is that described by Prudden, the specimens of which are in the medical museum of Columbia College, New York. There were over 1,182 distinct tumors distributed on the nerves of the body. These tumors rarely are painful, but may cause symptoms through pressure on neighboring structures.

**Generalized Neuro-fibromatosis: von Recklinghausen's Disease: Fibroma Molluscum.**—Special attention was directed to this particular form of multiple neuroma by von Recklinghausen in 1882. The disease presents several groups of lesions:

1. **CUTANEOUS.**—(a) Soft, fibrous nodules, some sessile, others pedunculated, varying in size and greatly in number, are scattered over the skin. They may increase in number as age advances. (b) Bluish spots, indicating atrophy of the corium where the fibromata are perforating. (c) Pigmentation, in the form of freckles, blotches, or diffuse areas. (d) Subcutaneous growths, at times of enormous size, causing the condition known as “elephantiasis neuromatosa.” Congenital nævi are frequent.

2. **NERVOUS.**—Tumors resembling plexiform neuromata may be present on any nerve trunk from the centre to the periphery. The variable situation leads to a variety of sensory or motor phenomena, especially as they may arise from the nerve roots within the spinal canal or cranium. Cases resembling tabes, syringomyelia and spastic paralysis are reported. The patients often show mental changes and disturbance of speech.

3. **BONE LESIONS.**—Changes similar to those of osteomalacia occur in about 7 per cent. of the cases.

Other features may be mentioned: Three generations have been affected, or two or three members of a family, or a mother and several children. The lesions may develop during pregnancy and disappear after delivery. Brickner, after whom this syndrome has been named, collected 16 cases. The tumors do not always disappear. Adrian reported a case with multiple myomata of the stomach. A sarcomatous change may occur in the central tumors, but not in the optic and olfactory nerves which have not the sheath of Schwann. There may be associated glioma or other brain tumor.

The nature of the disease is unknown. The occurrence of the pigmentation and the osteomalacia suggest an endocrine disturbance; but the familial and hereditary features point rather to an embryonic origin.

The prognosis depends on the possibility of successful removal of such tumors as are causing greatest inconvenience.

“**Tubercula dolorosa.**”—Multiple neuromata may especially affect the terminal cutaneous branches of the sensory nerves and lead to small subcutaneous painful nodules, often found on the face, breast, or about the joints. They may be associated with tumors of the nerve trunks.

“**Amputation Neuromata.**”—These bulbous swellings may form on the central ends of nerves which have been divided in injuries or operations. They are especially common after amputations. They are due to the tangled coil of axis-cylinder processes growing down from the central stump in an effort to reach their former end structures. They are very painful and usually require surgical removal but may recur.

### III. DISEASES OF THE CEREBRAL NERVES

#### OLFACTORY NERVES AND TRACTS

The functions of the olfactory nerves may be disturbed at their origin, in the nasal mucous membrane, at the bulb, in the course of the tract, or at the centres in the brain. The disturbances may be manifested in subjective sensations of smell, complete loss of the sense, and occasionally in hyperæsthesia.

**Subjective Sensations; Parosmia.**—Hallucinations of this kind are found in the insane and in epilepsy. The aura may be represented by an unpleasant odor, described as resembling chloride of lime, burning rags, or feathers. In a few cases with these subjective sensations tumors have been found in the hippocampi. In rare instances, after injury of the head, the sense is perverted—odors of the most different character may be alike, or the odor may be changed, as in a patient noted by Morell Mackenzie, who for some time could not touch cooked meat, as it smelt to her exactly like stinking fish.

**Increased sensitiveness (hyperosmia)** occurs chiefly in nervous, hysterical women, in whom it may sometimes be developed so greatly that, like a dog, they can recognize the difference between individuals by the odor alone.

**Anosmia; Loss of the Sense of Smell.**—This may be produced by: (a) Affections of the origin of the nerves in the mucous membrane, which is perhaps the most frequent cause. It is not uncommon with chronic nasal catarrh and polypi. In paralysis of the fifth nerve, the sense of smell may be lost on the affected side, owing to interference with secretion. It is doubtful whether the cases of loss of smell following the inhalations of foul or strong odors come under this or under the central division.

(b) Lesions of the bulbs or of the tracts. In falls or blows, in caries of the bones, and in meningitis or tumor, the bulbs or the olfactory tracts may be involved. After an injury to the head the loss of smell may be the only symptom. Mackenzie noted a case of a surgeon who was thrown from his gig and lighted on his head. The injury was slight, but the anosmia which followed was persistent. In tabes the sense of smell may be lost, possibly owing to atrophy of the nerves.

(c) Lesions of olfactory centres. There are congenital cases in which the structures have not developed. Cases have been reported in which anosmia has been associated with disease in the hemisphere.

To test the sense of smell the pungent bodies, such as ammonia, which act upon the fifth nerve, should not be used, but such substances as cloves, peppermint, and musk. This sense is readily tested by having two or three bottles containing the essential oils. In all instances a rhinoscopic examination should be made, as the condition may be due to local causes. The *treatment* is unsatisfactory even in the cases due to local nasal lesions.

## OPTIC NERVE AND TRACT

### (1) *Lesions of the Retina*

These are of importance to the physician, and information of the greatest value may be obtained by a systematic examination of the eye grounds. Only a brief reference can be made to the more important appearances.

**Retinitis.**—This occurs in certain general affections, more particularly in nephritis, syphilis, leukaemia, and anaemia. The common feature in all those is the occurrence of hæmorrhage and the development of opacities. There may also be a diffuse cloudiness due to effusion of serum. The hæmorrhages are in the layer of nerve fibres. They vary greatly in size and form, but often follow the course of vessels. When recent the color is bright red, but they gradually change and old hæmorrhages are almost black. The white spots are

due either to fibrinous exudate or to fatty degeneration of the retinal elements, and occasionally to accumulation of leucocytes or to a localized sclerosis of the retinal elements. The more important forms are:

**ALBUMINURIC RETINITIS** occurs in chronic nephritis, particularly in the interstitial form. The percentage of cases affected is from 15 to 25. There are instances in which these retinal changes occur at a stage when albuminuria may be slight or transient; but in such instances it will be found that there is a marked arterio-sclerosis. Gowers recognized a *degenerative* form (most common), in which, with the retinal changes, there may be scarcely any alteration in the disk; a *hæmorrhagic* form, with many hæmorrhages and but slight signs of inflammation; and an *inflammatory* form, in which there is much swelling of the retina and obscuration of the disk. It is noteworthy that in some instances the inflammation of the optic nerve predominates over the retinal changes, and one may be in doubt for a time whether the condition is associated with renal changes or dependent upon intracranial disease.

**SYPHILITIC RETINITIS**.—In the acquired form this is less common than choroiditis. In congenital syphilis *retinitis pigmentosa* is sometimes found.

**RETINITIS IN ANÆMIA**.—A patient may become blind after a large hæmorrhage, either suddenly or within two or three days, and in one or both eyes. Occasionally the loss may be permanent and complete. In some of these instances a neuro-retinitis has been found, probably sufficient to account for the symptoms. In the more chronic anæmias, particularly the pernicious form, retinitis is common, as determined first by Quincke.

In **MALARIA** retinitis or neuro-retinitis may be present, as noted by Stephen Mackenzie. It is seen only in the chronic cases with anæmia, and is not nearly so common proportionately as in pernicious anæmia.

**LEUKÆMIC RETINITIS**.—In this affection the retinal veins are large and distended; there is also a peculiar retinitis, as described by Liebreich. It is not very common. There are numerous hæmorrhages and white or yellow areas, which may be large and prominent. In one case the retina post mortem was dotted with many small, opaque, white spots, looking like little tumors, the larger of which had a diameter of nearly 2 mm.

Retinitis is also found occasionally in diabetes, in purpura, in chronic lead poisoning, and sometimes as an idiopathic affection.

**Functional Disturbances of Vision**.—(a) **TOXIC AMAUROSIS**.—This occurs in uræmia and may follow convulsions or come on independently. The condition, as a rule, persists only for a day or two. This form of amaurosis occurs in poisoning by lead, alcohol, and occasionally by quinine. It seems more probable that the poisons act on the centres and not on the retina.

(b) **TOBACCO AMBLYOPIA**.—The loss of sight is usually gradual, equal in both eyes, and affects particularly the centre of the field of vision. The eye-grounds may be normal, but occasionally there is congestion of the disks. On testing the color fields a central scotoma for red and green is found in all cases. Ultimately, if the use of tobacco is continued, organic changes may develop with atrophy of the disk.

(c) **HYSTERICAL AMAUROSIS**.—More frequently this is loss of acuteness of vision—amblyopia—but the loss of sight in one or both eyes may apparently be complete.

The condition will be mentioned under hysteria.

(d) **NIGHT-BLINDNESS—NYCTALOPIA**—the condition in which objects are clearly seen during the day or by strong artificial light, but become invisible in the shade or in twilight, and *hemeralopia*, in which objects can not be clearly seen without distress in daylight or in a strong artificial light, but are readily seen in a deep shade or in twilight, are rare functional anomalies which may occur in epidemic form.

(e) **RETINAL HYPERÆSTHESIA** is sometimes seen in hysterical women, but is not frequent in actual retinitis. It may occur with albuminuric retinitis and with aortic insufficiency.

## (2) Lesions of the Optic Nerve

**Optic Neuritis (Papillitis; Choked Disk).**—In the first stage there is congestion of the disk and the edges are blurred and striated. In the second stage the congestion is more marked; the swelling increases, the striation also is more visible. The physiological cupping disappears and hemorrhages are not uncommon. The arteries present little change, the veins are dilated, and the disk may swell greatly. In slight grades the swelling gradually subsides and occasionally the nerve recovers completely. In instances in which the swelling and exudate are very great the subsidence is slow, and when it finally disappears there is complete atrophy of the nerve. The retina may participate in the inflammation, which is then a neuro-retinitis.

This condition is of the greatest importance in diagnosis. It may exist in its early stages without any disturbance of vision, and even with extensive papillitis the sight may for a time be good.

Optic neuritis is seen occasionally in anæmia and lead poisoning, more commonly in nephritis as neuro-retinitis. It occurs occasionally as a primary idiopathic affection. The frequent connection with intracranial disease, particularly tumor, makes its presence of great value. The nature of the growth is without influence. In over 90 per cent. of such instances the choked disk is bilateral. It is also found in meningitis, either the tuberculous or the simple form. In meningitis the inflammation may extend down the nerve sheath. In tumor, however, it is probable that mechanical conditions, especially venous stasis, are alone responsible for the oedematous swelling. It often subsides very rapidly after decompression has been performed.

**Optic Atrophy.**—This may be: (a) A *primary* affection. There is an hereditary form, in which the disease has developed in all the males of a family shortly after puberty. A large number of the cases of primary atrophy are associated with spinal disease, particularly tabes. Other causes which are assigned are cold, diabetes, the specific fevers, methyl alcohol, and lead.

(b) *Secondary* atrophy results from cerebral diseases, pressure on the chiasma or on the nerves, or, most commonly of all, as a sequence of papillitis.

The ophthalmoscopic appearances are different in the cases of primary and secondary atrophy. In the former the disk has a gray tint, the edges are well defined, and the arteries look almost normal; whereas in the consecutive atrophy the disk has a staring opaque white aspect, with irregular outlines, and the arteries are very small.

The symptom of optic atrophy is loss of sight, proportionate to the damage in the nerve. The change is in three directions: "(1) Diminished acuity

vision; (2) alteration in the field of vision; and (3) altered perception of color (Gowers). The outlook in primary atrophy is bad.

### (3) *Affections of the Chiasma and Tract*

At the chiasma the optic nerves undergo partial decussation. Each optic tract, as it leaves the chiasma, contains nerve fibres which originate in the retina of both eyes. Thus, of the fibres of the right tract, part have come through the chiasma without decussating from the temporal half of the right retina, the other and larger portion of the fibres of the tract have decussated in the chiasma, coming as they do from the left optic nerve and the nasal half of the retina on the left side. The fibres which cross are in the middle portion of the chiasma, while the direct fibres are on each side. The following are the most important changes from lesions of the tract and chiasma:

**Unilateral Affection of Tract.**—If on the right side, this produces loss of function in the temporal half of the retina on the right side, and in the nasal half of the retina on the left side, so that there is only half vision, and the patient is blind to objects on the left side. This is termed homonymous hemianopia or lateral hemianopia. The fibres passing to the right half of each retina being involved, the patient is blind to objects in the left half of each visual field. The hemianopia may be partial and only a portion of the half field may be lost. The unaffected visual fields may have the normal extent, but in some instances there is considerable reduction. When the left half of one field and the right half of the other, or *vice versa*, are blind, the condition is known as heteronymous hemianopia.

**Disease of the Chiasma.**—(a) A lesion involves, as a rule, chiefly the central portion, in which the decussating fibres pass which supply the inner or nasal halves of the retina, producing in consequence loss of vision in the outer half of each field, or what is known as temporal hemianopia.

(b) If the lesion is more extensive it may involve not only the central portion, but also the direct fibres on one side of the commissure, in which case there is total blindness in one eye and temporal hemianopia in the other.

(c) Still more extensive disease is not infrequent from pressure of tumors in this region, the whole chiasma is involved, and total blindness results. The different stages in the process may often be traced in a single case from temporal hemianopia, then complete blindness in one eye with temporal hemianopia in the other, and finally complete blindness.

(d) A limited lesion of the outer part of the chiasma involves only the direct fibres passing to the temporal halves of the retina and inducing blindness in the nasal field, or, as it is called, nasal hemianopia. This, of course, is extremely rare. Double nasal hemianopia may occur as a manifestation of tabes and in tumors involving the outer fibres of each tract.

### (4) *Affections of the Tract and Centres*

The optic tract crosses the crus (cerebral peduncle) to the hinder part of the optic thalamus and divides into two portions, one of which (the lateral root) goes to the pulvinar of the thalamus, the lateral geniculate body, and to the anterior quadrigeminal body (superior colliculus). From these parts

in which the lateral root terminates, fibres pass into the posterior part of the internal capsule and enter the occipital lobe, forming the fibres of the optic radiation, which terminate in and about the cuneus, the region of the visual perceptive centre. It is held by some physiologists that the cortical visual centre is not confined to the occipital lobe alone, but embraces the occipito-angular region.

A lesion of the fibres of the optic path anywhere between the cortical centre and the chiasma will produce hemianopia. The lesion may be situated: (a) In the optic tract itself. (b) In the region of the thalamus, lateral geniculate body, and the corpora quadrigemina, into which the larger part of each tract enters. (c) A lesion of the fibre passing from the centre just mentioned to the occipital lobe. This may be either in the hinder part of the internal capsule or the white fibres of the optic radiation. (d) Lesion of the cuneus. Bilateral disease of the cuneus may result in total blindness. (e) There is clinical evidence to show that lesion of the angular gyrus may be associated with visual defect, not so often hemianopia as crossed amblyopia, dimness of vision in the opposite eye, and great contraction in the field of vision. Lesions in this region are associated with mind-blindness, a condition in which there is failure to recognize the nature of objects.

The effects of lesions in the optic nerve in different situations from the retinal expansion to the brain cortex are as follows: (1) Of the optic nerve, total blindness of the corresponding eye; (2) of the optic chiasma, either temporal hemianopia, if the central part alone is involved, or nasal hemianopia, if the lateral region of each chiasma is involved; (3) lesion of the optic tract between the chiasma and the lateral geniculate body produces lateral hemianopia; (4) lesion of the central fibres of the nerve between the geniculate bodies and the cerebral cortex produces lateral hemianopia; (5) lesion of the cuneus causes lateral hemianopia; and (6) lesion of the angular gyrus may be associated with hemianopia, sometimes crossed amblyopia, and the condition known as mind-blindness. (See Fig. 23.)

**Diagnosis of Lesions of the Optic Nerve and Tract.**—This depends much on the associated symptoms. The *significance* of hemianopia varies. There is a functional hemianopia associated with migraine and hysteria. In a considerable proportion of all cases there are signs of organic brain disease. In a certain number of instances of slight lesions of the occipital lobe hemichromatopsia has been observed. The homonymous halves of the retina as far as the fixation point are dulled, or blind for colors. Hemiplegia is common, in which event the loss of power and blindness are on the same side. Thus, a lesion in the left hemisphere involving the motor tract produces right hemiplegia, and when the fibres of the optic radiation are involved in the internal capsule there is also lateral hemianopia, so that objects in the field of vision to the right are not perceived. Hemianæsthesia is not uncommon in such cases, owing to the close association of the sensory and visual tracts at the posterior part of the internal capsule. Certain forms of aphasia also occur in many of the cases.

#### MOTOR NERVES OF THE EYEBALL

**Third Nerve** (*Nervus oculomotorius*).—The nucleus of origin of this nerve is situated in the floor of the aqueduct of Sylvius; the nerve passes through

the crus at the side of which it emerges. Passing along the wall of the cavernous sinus, it enters the orbit through the sphenoidal fissure and supplies, by its superior branch, the levator palpebræ superioris and the superior rectus,

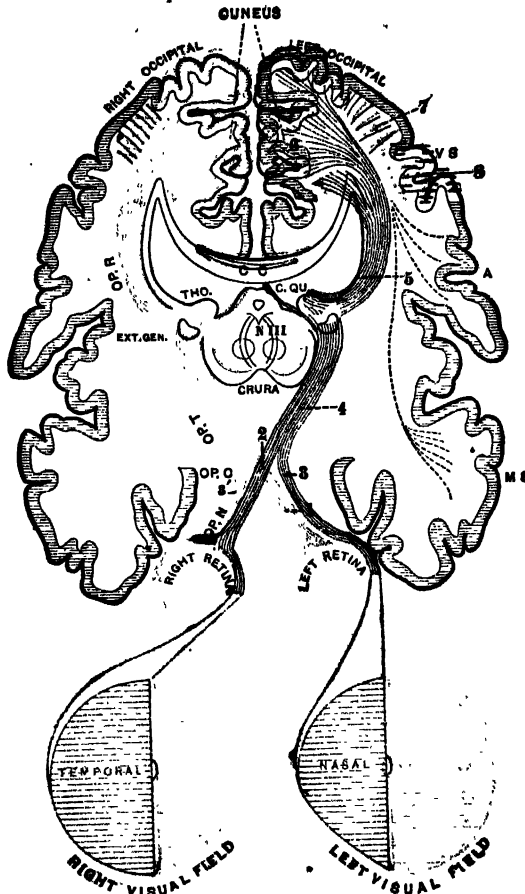


FIG. 23.—DIAGRAM OF VISUAL PATHS. (From Violet, modified.)

OP. N., Optic nerve. OP. C., Optic chiasm. OP. T., Optic tract. OP. R., Optic radiations. EXT. GEN., External geniculate body. THO., Optic thalamus. C. QU., Corpora quadrigemina. C. C., Corpus callosum. V. S., Visual speech centre. A. S., Auditory speech centre. H. S., Motor speech centre. A lesion at 1 causes blindness of that eye; at 2, bi-temporal hemianopia; at 3, nasal hemianopia. Symmetrical lesions at 3 and 3' would cause bi-nasal hemianopia; at 4, hemianopia of both eyes, with hemianopic pupillary inaction; at 5 and 6, hemianopia of both eyes, pupillary reflexes normal; at 7, amblyopia, especially of opposite eye; at 8, on left side, word-blindness.

and by its inferior branch the internal and inferior recti muscles and the inferior oblique. Branches pass to the ciliary muscle and the constrictor of the iris. Lesions may affect the nucleus of the nerve in its course and cause either paralysis or spasm.

**PARALYSIS.**—A nuclear lesion is usually associated with disease of the centres for the other eye muscles, producing general ophthalmoplegia. More

commonly the nerve itself is involved in its course, either by meningitis, gum-mata, or aneurism, or is attacked by a neuritis, as in diphtheria. Complete paralysis is accompanied by the following symptoms:

Paralysis of all the muscles, except the superior oblique and external rectus, by which the eye can be moved outward and a little downward and inward. There is divergent strabismus. There is *ptosis* or drooping of the upper eyelid, owing to paralysis of the levator palpebræ. The *pupil* is usually dilated. It does not contract to light, and the power of accommodation is lost. The most striking features of this paralysis are the external strabismus, with diplopia or double vision, and the ptosis. In very many cases the affection of the third nerve is partial. Thus the levator palpebræ and the superior rectus may be involved together, or the ciliary muscles and the iris may be affected and the external muscles may escape.

There is a remarkable form of *recurring* oculo-motor paralysis affecting chiefly women, and involving all the branches of the nerve. In some cases the attacks have come on at intervals of a month; in others a much longer period has elapsed. The attacks may persist throughout life. They are sometimes associated with pain in the head and sometimes with migraine. Mary Sherwood collected 23 cases from the literature.

Ptosis is a common and important sign in nervous affections. We may here briefly refer to the conditions under which it may occur: (a) A congenital, incurable form; (b) the form associated with definite lesion of the third nerve, either in its course or at its nucleus. This may come on with paralysis of the superior rectus alone or with paralysis of the internal and inferior recti as well. (c) There are instances of complete or partial ptosis associated with cerebral lesions without any other branch of the third nerve being paralyzed. (d) Hysterical ptosis, which is double and occurs with other hysterical symptoms. (e) Pseudo-ptosis, due to affection of the sympathetic nerve, is associated with symptoms of vaso-motor palsy, such as elevation of the temperature on the affected side with redness and œdema of the skin. Contraction of the pupil exists on the same side and the eyeball appears rather to have shrunk into the orbit. (f) In idiopathic muscular atrophy, when the face muscles are involved, there may be marked bilateral ptosis. And, lastly, in weak, delicate women there may be a transient ptosis, particularly in the morning.

Among the most important of the symptoms of the third-nerve paralysis are those which relate to the ciliary muscle and iris.

CYCLOPLEGIA, paralysis of the ciliary muscle, causes loss of the power of accommodation. Distant vision is clear, but near objects cannot be properly seen. In consequence the vision is indistinct, but can be restored by the use of convex glasses. This may occur in one or in both eyes; in the latter case it is usually associated with disease in the nuclei. Cycloplegia is an early and frequent sign in diphtheritic paralysis and occurs also in tabes.

IRIDOPLEGIA, or paralysis of the iris, occurs in three forms (Gowers):

(a) *Accommodation iridoplegia*, in which the pupil does not alter in size during the act of accommodation. To test this the patient should look first at a distant and then at a near object in the same line of vision.

(b) *Reflex Iridoplegia*.—The path for the iris reflex is along the optic nerve and tract to its termination, then to the nucleus of the third nerve,

and along the trunk of this nerve to the ciliary ganglion, and so through the ciliary nerves to the eyes. Each eye should be tested separately, the other one being covered. The patient should look at a distant object in a dark part of the room; then a light is brought suddenly in front of the eye at a distance of three or four feet, so as to avoid the effect of accommodation. Loss of this iris reflex with retention of the accommodation contraction is known as the Argyll-Robertson pupil.

(c) *Loss of the Skin Reflex.*—If the skin of the neck is pinched or pricked the pupil dilates reflexly, the afferent impulses being conveyed along the cervical sympathetic. Erb pointed out that this skin reflex is lost usually in association with the reflex contraction, but the two are not necessarily conjoined. In iridoplegia the pupils are often small, particularly in spinal disease, as in the characteristic small pupils of tabes—spinal myosis. Iridoplegia may coexist with a pupil of medium size.

Inequality of the pupils—*anisocoria*—is not infrequent in syphilis, paresis and tabes. It may occur in perfectly healthy individuals.

**SPASM.**—Occasionally in meningitis and in hysteria there is spasm of the muscles supplied by the third nerve, particularly the internal rectus and the levator palpebræ. *Nystagmus* is a rhythmical contraction of the eye muscles met with in many congenital and acquired lesions of the brain, particularly in multiple sclerosis. It may be hereditary and has been traced through four generations in association with head nodding (Yawger). Lid nystagmus may also be present. It is met with in albinos. The nystagmus of miners is apparently due to poor light.

**Fourth Nerve** (*Nervus trochlearis*).—This supplies the superior oblique muscle. In its course around the outer surface of the crus and in its passage into the orbit it is liable to be compressed by tumors, by aneurism, or in the exudation of basilar meningitis. Its nucleus in the upper part of the fourth ventricle may be involved by tumors or undergo degeneration with the other ocular nuclei. The superior oblique muscle acts in such a way as to direct the eyeball downward and rotate it slightly. The paralysis causes defective downward and inward movement, often too slight to be noticed. The head is inclined somewhat forward and toward the sound side, and there is double vision when the patient looks down.

**Sixth Nerve.**—Emerging at the junction of the pons and medulla, it passes forward in a long and exposed course to the orbit, and supplies the external rectus muscle. It is often involved in meningeal exudate, compressed by tumors and possibly involved in an independent neuritis. When paralyzed, there is internal squint with diplopia on attempting to look outwards. The true and the false images are parallel, and grow further apart on looking to the paralyzed side. When the nucleus is involved, the internal rectus of the opposite eye may be paralyzed as the nucleus sends fibres up in the pons to that part of the nucleus of the opposite third nerve which supplies the internal rectus. In one symptom-complex there is a combination of otitis media with complete paralysis of the sixth nerve. The inflammation travels to the apex of the petrous bone; then to the sixth nerve. The outlook is usually good.

**General Features of Paralysis of the Motor Nerves of the Eye.**—Gowers divided them into five groups:

(a) *Limitation of Movement.*—Thus, in paralysis of the external rectus, the eyeball can not be moved outward. When the paralysis is incomplete the movement is deficient in proportion to the degree of the palsy.

(b) *Strabismus.*—The axes of the eyes do not correspond. Thus, paralysis of the internal rectus causes a divergent squint; of the external rectus, a convergent squint. At first this is evident only when the eyes are moved in the direction of the action of the weak muscle. The deviation of the axis of the affected eye from parallelism with the other is called the primary deviation.

(c) *Secondary Deviation.*—If, while the patient is looking at an object, the sound eye is covered, so that he fixes the object looked at with the affected eye only, the sound eye is moved still further in the same direction—e. g., outward, when there is paralysis of the opposite internal rectus. This is known as secondary deviation. It depends upon the fact that, if two muscles are acting together, when one is weak and an effort is made to contract it, the increased effort—innervation—acts powerfully upon the other muscle, causing an increased contraction.

(d) *Erroneous Projection.*—"We judge of the relation of external objects to each other by the relation of their images on the retina; but we judge of their relation to our own body by the position of the eyeball as indicated to us by the innervation we give to the ocular muscles" (Gowers). With the eyes at rest in the mid-position, an object at which we are looking is directly opposite our face. Turning the eyes to one side, we recognize that object in the middle of the field or to the side of this former position. We estimate the degree by the amount of movement of the eyes, and when the object moves and we follow it we judge of its position by the amount of movement of the eyeballs. When one ocular muscle is weak the increased innervation gives the impression of a greater movement of the eye than has really taken place. The mind, at the same time, receives the idea that the object is further on one side than it really is, and in an attempt to touch it the finger may go beyond it. As the equilibrium of the body is in a large part maintained by a knowledge of the relation of external objects to it obtained by the action of the eye muscles, this erroneous projection disturbs the harmony of these visual impressions and may lead to giddiness—*ocular vertigo*.

(e) *Double Vision.*—This is one of the most disturbing features of paralysis of the eye muscles. The visual axes do not correspond, so that there is a double image—*diplopia*. That seen by the sound eye is termed the true image; that by the paralyzed eye, the false. In simple or homonymous diplopia the false image is "on the same side of the other as the eye by which it is seen." In crossed diplopia it is on the other side. In convergent squint the diplopia is simple; in divergent it is crossed.

**Sympathetic Fibres.**—These supply the muscle of Müller, which pushes the eye forward, fibres in the muscle elevating the upper lid and the dilator muscle of the pupil. Stimulation causes widening of the palpebral fissure, exophthalmus and a dilated pupil, as in Graves' disease. Paralysis causes a slight degree of ptosis, enophthalmus and a contracted pupil.

**Ophthalmoplegia.**—Two forms are recognized—*ophthalmoplegia externa* and *interna* which may occur separately or together.

**OPHTHALMOPLÉGIA EXTERNA.**—The condition is one of more or less complete palsy of the external muscles of the eyeball, due usually to a slow degen-

eration in the nuclei of the nerves, but sometimes to pressure of tumors or to basilar meningitis. It is often, but not necessarily, associated with ophthalmoplegia interna. Of 62 cases analyzed by Siemerling in only 11 could syphilis be positively determined. The levator muscles of the eyelids and the superior recti are first involved, and gradually the other muscles, so that the eyeballs are fixed and the eyelids droop. There is sometimes slight protrusion of the eyeballs. The disease is essentially chronic and may last for years. It is found particularly in association with general paresis, tabes, and in progressive muscular atrophy. Mental disorders were present in 11 of the 62 cases. With it may be associated optic atrophy and affections of other cerebral nerves. Occasionally, as noted by Bristowe, it may be functional.

**OPHTHALMOPLÉGIA INTERNA.**—Jonathan Hutchinson applied this term to a progressive paralysis of the internal ocular muscles, causing loss of pupillary action and the power of accommodation. When the internal and external muscles are involved the affection is known as total ophthalmoplegia, and in a majority of the cases the two conditions are associated. In some instances the internal form may depend upon disease of the ciliary ganglion.

While, as a rule, ophthalmoplegia is a chronic process, an acute form may be associated with encephalitis or hemorrhagic softening of the nuclei of the ocular nerves. There is usually marked cerebral disturbance in the latter.

**Treatment of Ocular Palsies.**—It is important to ascertain the cause. The forms associated with tabes are obstinate and resist treatment. Occasionally a palsy, complete or partial, may pass away spontaneously. The cases associated with chronic degenerative changes, as in paresis and bulbar paralysis, are little affected by treatment. On the other hand, in syphilitic cases, specific treatment is often beneficial. Arsenic and strychnia, the latter hypodermically, may be employed. In any case in which the onset is acute with pain, hot fomentations and counter-irritation or leeches applied to the temple give relief. Treatment by electricity has been employed without any special effect. The diplopia may be relieved by the use of prisms, or it may be necessary to cover the affected eye with an opaque glass.

## FIFTH NERVE

(*Nervus trigeminus*)

**Etiology.**—Paralysis may result from: (a) Disease of the pons, particularly hemorrhage, tumor or sclerosis. (b) Injury or disease at the base of the brain. Fracture rarely involves the nerve but meningitis, acute or chronic, and caries of the bone are not uncommon causes. (c) The branches may be affected as they pass out—the first division by tumors pressing on the cavernous sinus or by aneurism; the second and third divisions by growths in the sphenomaxillary fossa. (d) Primary neuritis, which is rare.

**Symptoms.**—(a) **SENSORY PORTION.**—Disease of the fifth nerve may cause loss of sensation in the parts supplied, including the half of the face, the corresponding side of the head, the conjunctiva, the mucosa of the lips, tongue, hard and soft palate, and of the nose of the same side. The anæsthesia may be preceded by tingling or pain. The muscles of the face are also insensible and the movements may be slower. The sense of smell is interfered with,

owing to dryness of the mucous membrane. The salivary, lachrymal, and buccal secretions may be lessened, and the teeth may become loose. Unless properly guarded from injury an ulcerative inflammation of the eye may follow. This was supposed to be due to nutritional changes from paralysis of so-called trophic nerve fibres. This idea has been overthrown by the large number of cases in which the Gasserian ganglion has been removed without consequent inflammation of the eye. *Herpes* may occur in the region supplied by the nerve, usually the upper branch, and is associated with much pain, which may last for months or years. In herpes zoster with the neuritis there may be slight enlargement of the cervical glands. (See Trigeminal Neuralgia.)

(b) MOTOR PORTION.—The inability to use the muscles of mastication on the affected side is the distinguishing feature of paralysis of this portion of the nerve. It is recognized by placing the finger on the masseter and temporal muscles, and, when the patient closes the jaw, the feebleness of their contraction is noted. If paralyzed, the external pterygoid can not move the jaw toward the unaffected side; and when depressed, the jaw deviates to the paralyzed side. Motor paralysis of the fifth nerve results from lesions of the nucleus or in the peripheral part of the motor division.

*Spasm of the Muscles of Mastication.*—*Trismus*, the masticatory spasm of Romberg, may be tonic or clonic, and is either an association phenomenon in general convulsions or, more rarely, an independent affection. In the tonic form the jaws are kept close together—lock-jaw—or can be separated only for a short space. The muscles of mastication can be seen in contraction and felt to be hard; the spasm is often painful. This tonic contraction is an early symptom in tetanus, and is sometimes seen in tetany. A form of this tonic spasm occurs in hysteria. Occasionally trismus follows exposure to cold, and is said to be due to reflex irritation from the teeth, the mouth, or caries of the jaw. It may also be a symptom of organic disease due to irritation near the motor nucleus of the fifth nerve.

*Clonic spasm of the muscles supplied by the fifth occurs in the form of rapidly repeated contractions, as in "chattering teeth."* This is rare apart from general conditions, though cases are on record, usually in women late in life, in whom this isolated clonic spasm of the muscles of the jaw has been found. In another form of clonic spasm sometimes seen in chorea there are forcible single contractions.

(c) GUSTATORY.—There are two views concerning the course of the fibres that carry gustatory impulse from this part of the tongue. According to some they take a devious path, passing with the chorda tympani to the geniculate ganglion, thence by the great superficial petrosal nerve to Meckel's ganglion, and this they leave to reach the maxillary nerve, which they follow through the trigeminal nerve to the brain. It seems more probable that the fibres pass with the facial nerve to the brain directly from the geniculate ganglion by the nervus intermedius of Wrisberg. The studies of Cushing upon patients who have had the Gasserian ganglion removed show that the sense of taste is retained.

The *diagnosis* of disease of the trifacial nerve is rarely difficult. It must be remembered that the preliminary pain and hyperæsthesia are sometimes mistaken for ordinary neuralgia. The loss of sensation and the palsy of the

muscles of mastication are readily determined. Involvement of one branch means a lesion peripheral to the Gasserian ganglion.

**Treatment.**—When the pain is severe morphia may be required and local applications are useful. If there is a suspicion of syphilis, appropriate treatment should be given. Faradization is sometimes beneficial.

## FACIAL NERVE

**Paralysis (Bell's Palsy).**—**ETIOLOGY.**—The facial or seventh may be paralyzed by (a) lesions of the cortex—supranuclear palsy; (b) lesions of the nucleus itself; or (c) involvement of the nerve trunk in its tortuous course within the pons and through the wall of the skull.

(a) *Supranuclear paralysis.* due to lesion of the cortex or of the facial fibres in the corona radiata or internal capsule, is, as a rule, associated with hemiplegia. It may be due to tumors, abscess, inflammation, or softening in the cortex or in the region of the internal capsule. It is distinguished from the peripheral form by the persistence of the normal electrical excitability of both nerves and muscles and the frequent absence of involvement of the upper branches of the nerve, so that the orbicularis palpebrarum, frontalis, and corrugator muscles are spared. In rare instances these muscles are paralyzed. In this form the voluntary movements are more impaired than the emotional. Isolated paralysis—monoplegia facialis—due to involvement of the cortex or of the fibres in their path to the nucleus, is uncommon. In the majority of cases supranuclear facial paralysis is part of a hemiplegia. Paralysis is on the same side as that of the arm and leg because the facial muscles bear the same relation to the cortex as the spinal muscles. The nuclei of origin on either side of the middle line in the medulla are united by decussating fibres with the cortical centre on the opposite side. (See Fig. 22.) A few fibres reach the nucleus from the cerebral cortex of the same side, and this uncrossed path may innervate the upper facial muscles.

(b) The *nuclear paralysis* caused by lesions of the nerve centres in the medulla is seen in tumors, encephalitis, chronic softening, and hemorrhage. It may be involved in polio-myelitis. In diphtheria this centre may also be attacked. The symptoms are practically similar to those of an affection of the nerve fibre itself—infranuclear paralysis.

(c) *Involvement of the Nerve Trunk.*—Paralysis may result from:

(1) Involvement of the nerve as it passes through the pons—that is, between its nucleus in the floor of the fourth ventricle and the point of emergence in the postero-lateral aspect of the pons. The specially interesting feature in connection with involvement of this part is the production of what is called alternating or *crossed paralysis*, the face being involved on the same side as the lesion, and the arm and leg on the opposite side, since the motor path is involved above the point of decussation in the medulla (Fig. 22). This occurs only when the lesion is in the lower section of the pons. A lesion in the upper half of the pons involves the fibres not of the outgoing nerve on the same side, but of the fibres from the hemispheres before they have crossed to the nucleus of the opposite side. In this case there would be paralysis of the face and limbs on the side opposite to the lesion. The palsy would resemble the cerebral form, involving only the lower fibres of the facial nerve.

(2) The nerve may be involved at its point of emergence by tumors, particularly by the cerebello-pontine growths, by gummata, meningitis, or occasionally it may be injured in fracture of the base.

(3) In passing through the Fallopian canal the nerve may be involved in disease of the ear, particularly by caries of the bone in otitis media. This is a common cause. Fibrositis about the nerve may be responsible.

(4) As the nerve emerges from the styloid foramen it is exposed to injuries and blows which may cause paralysis. The fibres may be cut in the removal of tumors in this region, or the paralysis may be caused by pressure of the forceps in an instrumental delivery.

(5) Exposure to cold is a common cause, inducing a neuritis of the nerve within the Fallopian canal. Reik believes that in most of these cases there is an acute otitis media from which the nerve is involved.

(6) Syphilis is not an infrequent cause, and the paralysis may appear early with the secondary symptoms.

(7) It may occur in association with herpes.

*Facial diplegia* is a rare condition occasionally found in affections at the base of the brain, lesions in the pons, simultaneous involvement of the nerves in ear-disease, and in diphtheritic paralysis. Disease of the nuclei or symmetrical involvement of the cortex might also produce it. It may occur as a congenital affection. H. M. Thomas described two cases in one family.

**SYMPTOMS.**—In the peripheral facial paralysis all the branches of the nerve are involved. The face on the affected side is immobile and can neither be moved at will nor participate in any emotional movements. The skin is smooth and the wrinkles are effaced, a point particularly noticeable on the forehead of elderly persons. The eye can not be closed or the forehead wrinkled, the lower lid droops, and the eye waters. On the affected side the angle of the mouth is lowered, and in drinking the lips are not kept in close apposition to the glass, so that the liquid is apt to run out. In smiling or laughing the contrast is most striking, as the affected side does not move, giving a curious unequal appearance to the two sides of the face. In long standing cases, when the reaction of degeneration is present, if the patient tries to close the eyes while looking fixedly at an object the lids on the sound side close firmly, but on the paralyzed side there is only a slight inhibitory droop of the upper lid, and the eye is turned upward and outward by the inferior oblique. On asking the patient to show his upper teeth, the angle of the mouth is not raised. In all these movements the face is drawn to the sound side by the action of the muscles. Speaking may be slightly interfered with, owing to the imperfection in the formation of the labial sounds. Whistling can not be performed. In chewing the food, owing to the paralysis of the buccinator, particles collect on the affected side. The paralysis of the nasal muscles is seen on asking the patient to sniff. Owing to the fact that the lips are drawn to the sound side, the tongue, when protruded, looks as if it were pushed to the paralyzed side; but on taking its position from the incisor teeth, it will be found to be in the middle line. The reflex movements are lost in this peripheral form.

The *sensory functions* of the facial nerve, to which much attention has been paid by Cushing, Mills and others, are ministered to by the geniculate ganglion, the intermediary nerve of Wrisberg, and the chorda tympani, which

last has chiefly gustatory functions. It seems likely that deep sensibility with sense of pressure, position and passive movement runs in a separate afferent system in the motor nerve of the face. Cutaneous sensibility, both epicritic, by which we localize light touch, and protopathic, by which we recognize degrees of heat and cold, is not ministered to by the facial nerve proper. There are observations that would indicate, however, that the anterior part of the tongue and possibly a little strip of the skin of the auricle have a vestigial supply from this nerve.

When the nerve is involved after the chorda tympani has joined it, the sense of taste is lost in the anterior part of the tongue on the affected side. When the nerve is damaged outside the skull the sense of taste is unaffected. Hearing is often impaired in facial paralysis, most commonly by preceding ear disease. The paralysis of the stapedius muscle may lead to increased sensitiveness to musical notes. Herpes in the ear and in the distribution of the fifth nerve is sometimes associated with facial paralysis. Severe pain may precede or accompany the paralysis. It is usually in the ear and mastoid region but may radiate to the occipital and trigeminal distribution. The face on the affected side may be swollen.

The *electrical reactions*, which are those of a peripheral palsy, have considerable importance from a prognostic standpoint. Erb's rules are as follows: If there is no change, either faradic or galvanic, the prognosis is good and recovery takes place in from fourteen to twenty days. If the faradic and galvanic excitability of the nerve is only lessened and that of the muscle increased to the galvanic current and the contraction formula altered (the contraction sluggish  $AC < KC$ ), the outlook is relatively good and recovery will probably take place in from four to six weeks; occasionally in from eight to ten. When the reaction of degeneration is present and the mechanical excitability is altered, the prognosis is relatively unfavorable and recovery may not occur for two, six, eight, or even fifteen months.

**COURSE.**—This is usually favorable in the common form but not in those due to intracranial lesions or caries of bone. The onset in the form following cold is very rapid, developing perhaps within twenty-four hours, but rarely is the paralysis permanent. Hunt has drawn special attention to *recurrent* facial paralysis which may be on one or alternate sides—"relapsing alternating." In some instances contracture develops as the voluntary power returns, and the natural folds and the wrinkles on the affected side may be deepened, so that on looking at the face one at first may have the impression that the affected side is the sound one. This is corrected at once on asking the patient to smile, when it is seen which side of the face has the more active movement. Aretæus noted the difficulty sometimes experienced in determining which side was affected until the patient spoke or laughed.

**PERMANENT FACIAL PARALYSIS.**—One of the distressing sequels is permanent loss of power with immobility and the disfigurement resulting from the overaction of the muscles on the sound side. There are three groups of cases: (1) Those due to *trauma*, especially the birth palsies from injury by forceps. (2) Due to suppurative middle-ear disease, following scarlet fever, diphtheria, or sepsis of any kind, such as puerperal fever. (3) In a few cases following the ordinary Bell's paralysis. Even when paralysis exists from childhood, there may be slight voluntary control, and the muscles may respond

to faradic stimulus. The facial nerve in reality may have recovered or regenerated, and the disfigurement and loss of function result from the over-stretching of the degenerated muscles by the action of their opponents on the sound side (Turrell).

**DIAGNOSIS.**—This is usually easy. The distinction between the peripheral and central form is based on facts already mentioned.

**TREATMENT.**—In the cases which result from cold and are probably due to neuritis within the bony canal, hot applications should be made; subsequently counter-irritation by iodine or small blisters should be used. If the ear is diseased, free discharge should be obtained. The galvanic current may be employed to keep up the nutrition of the muscles. The positive pole should be placed behind the ear, the negative one along the zygomatic and other muscles. The application can be made daily for a quarter of an hour and the patient can make it himself. Massage in the course of the nerve and of the muscles of the face is also useful. A course of iodide of potassium may be given even when there is no indication of syphilis.

In those cases in which the nerve has been destroyed by an injury, during an operation or from disease, and when there has been no evidence of returning function after treatment for a few months, a nerve anastomosis should be performed. For this purpose either the spinal accessory or the hypoglossal nerve may be used. Though the normal conditions may never be completely regained, the motor power will be largely restored and the deformity lessened. This procedure, based on the results of physiological experimentation, makes one of the most striking of modern operations.

**Spasm.**—This may be limited to a few or involve all the muscles innervated by the facial nerve, and may be unilateral or bilateral. It is known also as *mimic spasm* or *convulsive tic*. Several different affections are usually considered under the name of facial spasm, but we speak here only of the simple spasm of the facial muscles, either primary or following paralysis, and do not include the habit spasm in children, or the *tic convulsif* of the French.

Gowers recognized two classes—one in which there is an organic lesion, and an idiopathic form. It is thought to be due also to reflex causes, such as the irritation from carious teeth. The disease usually occurs in adults, whereas the habit spasm and the *tic convulsif* of the French, often confounded with it, are most common in children. True mimic spasm occasionally comes on in childhood and persists. When the result of organic disease, there has usually been a lesion of the centre in the cortex, as in the case reported by Berkley, or pressure on the nerve at the base of the brain by aneurism or tumor.

**SYMPTOMS.**—The spasm may involve only the muscles around the eyes—blepharospasm—in which case there is constant, rapid, quick action of the orbicularis palpebrarum, which, in association with photophobia, may be tonic in character. More commonly the spasm affects the lateral facial muscles with those of the eye, and there is constant twitching of the side of the face with partial closure of the eye. The frontalis is rarely involved. In aggravated cases the depressors of the angle of the mouth, the levator menti, and the platysma myoides are affected. This spasm is confined to one side of the face in a majority of cases, though it may extend and become bilateral. It is increased by emotional causes and by voluntary movements of the face. As a rule, it is painless, but there may be tender points over the course of the fifth

nerve, particularly the supraorbital branch. Tonic spasm of the facial muscle may follow paralysis, and is said to result occasionally from cold.

The outlook in facial spasm is always dubious. A majority of the cases persist for years and are incurable.

**TREATMENT.**—Sources of irritation should be looked for and removed. When a painful spot is present, blistering or the application of the cauter may relieve it. The salicylates, mercury and iodide have seemed to help some patients and should be tried. Injection of alcohol (50 per cent.) into the facial nerve has been successful (Patrick). The needle should be introduced with great care and the alcohol injected very slowly until the first signs of weakness of the facial muscles appear. Weir Mitchell recommended freezing the cheek for a few minutes daily or every second day with the spray, and this, in some instances, is beneficial. Often the relief is transient and at every clinic patients may be seen who have run the gamut of all measures without material improvement. Severe cases may require surgical interference. The nerve may be divided near the stylomastoid foramen and an anastomosis made between it and the spinal accessory.

#### AUDITORY NERVE

The eighth nerve in reality consists of two separate nerves—the cochlear and vestibular roots. These two roots have entirely different functions, and are therefore best considered separately. The cochlear nerve is the one connected with the organ of Corti, and is concerned in hearing. The vestibular nerve is connected with the vestibule and semicircular canals, and has to do with the maintenance of equilibrium.

##### *The Cochlear Nerve*

The cortical centre for hearing is in the temporo-sphenoidal lobe. Primary disease of the auditory nerve in its centre or intracranial course is uncommon. More frequently the terminal branches are affected within the labyrinth. The ears are represented bilaterally in the brain and so a lesion on one side does not cause any marked disturbance, but if the left side is involved this may result in word-deafness. A tumor involving the region of the posterior quadrigeminal bodies causes deafness on the same side.

**Lesions of the nerve at the base** of the brain may result from the pressure of tumors, including the acoustic tumor, meningitis, hemorrhage, or traumatism. A primary degeneration of the nerve may occur in tabes. Primary disease of the terminal nuclei of the cochlear nerve is rare. A form results from epidemic cerebro-spinal meningitis, in which the nerve is frequently involved, causing permanent deafness. In young children the condition results in deafmutism.

**Internal Ear.**—In a majority of cases with auditory nerve symptoms the lesion is in the internal ear, either primary or the result of extension of disease of the middle ear. Two groups of symptoms may be produced—hyperæsthesia and irritation, and diminished function or nervous deafness.

(a) **HYPERÆSTHESIA AND IRRITATION.**—This may be due to altered function of the centre as well as of the nerve ending. True hyperæsthesia—hyper-

acusis—is a condition in which sounds, sometimes even those inaudible to other persons, are heard with great intensity. It occurs in hysteria and occasionally in cerebral disease. In paralysis of the stapedius low notes may be heard with intensity. In dysæsthesia, or dysacusis, ordinary sounds cause an unpleasant sensation, as commonly happens in connection with headache, when ordinary noises are badly borne.

*Tinnitus aurium* is a term employed to designate certain subjective sensations of ringing, roaring, ticking and whirring noises in the ear. It is a very common and often a distressing symptom. Certain drugs, such as quinine, may cause it. It is associated with many forms of ear disease and may result from pressure of wax on the drum. It is rare in organic disease of the central connections of the nerve. Sudden intense stimulation of the nerve may cause it. A not uncommon form is that in which the patient hears a continual *bruit* in the ear, and the noise has a systolic intensification, usually on one side. It may suggest an internal aneurism. A systolic murmur may be heard occasionally on auscultation in anæmia and neurasthenia. Subjective noises in the ear may precede an epileptic seizure and are sometimes present in migraine. In whatever form tinnitus exists, though slight and often regarded as trivial, it occasions great annoyance and mental distress, and has driven patients to suicide.

The *diagnosis* is readily made; but it is often extremely difficult to determine the cause of the tinnitus. The relief of constitutional states, such as anæmia or neurasthenia may result in cure. A careful local examination of the ear should always be made. One of the most worrying forms is the constant clicking, sometimes audible many feet away from the patient, and due probably to clonic spasm of the muscles connected with the Eustachian tube or of the levator palati. The condition may persist for years unchanged, and then disappear suddenly. The pulsating forms of tinnitus, in which the sound is like that of a systolic murmur, are almost invariably subjective. It is to be remembered that in children there is a systolic brain murmur, best heard over the ear, and in some instances appreciable in the adult.

(b) DIMINISHED FUNCTION OR NERVOUS DEAFNESS.—In testing for nervous deafness, if the tuning fork can not be heard when placed near the meatus, but the vibrations are audible by placing the foot of the tuning fork against the temporal bone, the conclusion may be drawn that the deafness is not due to involvement of the nerve. The vibrations are conveyed through the temporal bone to the cochlea and vestibule. The watch may be used for the same purpose, and if the meatus is closed and the watch is heard better in contact with the mastoid process than when opposite the open meatus, the deafness is probably not nervous. Disturbance of the function of the auditory nerve is not a very frequent symptom in brain disease, but in all cases the function of the nerve should be carefully tested.

#### *The Vestibular Nerve*

Our sense of position in space and the control of the balance of the body depend partly on proper function of the vestibular nerve and its central associations in the cerebellum and cerebrum. The paths from the labyrinth are

not accurately known, beyond the group of Deiters' nuclei; there is uncertainty, both as to tracts and centres.

Disturbance of the relation of the body to space, or of its balance, produces in consciousness the unpleasant sensation which we call dizziness or vertigo. It results from a discord between the impressions arising in the labyrinth, the cerebellum, the eye muscles and elsewhere, and a failure to coördinate these in the cerebrum. The controlling factor is the vestibular mechanism.\* The cochlear nerves are often involved simultaneously, producing tinnitus, and nystagmus may be associated.

An apprehension, not a true vertigo, is common in looking from a height, especially in those with disease of the internal ear. True vertigo is always accompanied by a sensation of falling or turning, even when the person is in bed, and if standing, there is inco-ordination of the muscles, with staggering or falling. The patient may feel that he is moving or the objects about him appear to rotate. The direction in which he falls is variable and of special importance. Nystagmus is often associated and the direction and intensity should be studied.

**(1) Auditory (Labyrinthine) Vertigo—Ménière's Syndrome.**—In 1861 Ménière described an affection characterized by noises in the ear, attacks of vertigo which may be associated with vomiting and loss of consciousness; in many cases there is progressive loss of hearing. Bárány groups the conditions in which the labyrinth may be affected and vertigo occur under the following heads: (a) Acute infectious diseases, influenza, cerebro-spinal meningitis, etc. (b) Chronic infectious diseases, syphilis particularly. (c) Constitutional conditions and intoxications. Hæmorrhage into the labyrinth (in leukaemia, purpura hæmorrhagica, pernicious anæmia); chlorosis, thyroid intoxications, arterio-sclerosis, etc. (d) Tumors and diseases of the central nervous system; tumors of the acoustic nerve, cerebellum, pons, and fourth ventricle, meningitis, cerebellar abscess, multiple sclerosis, tabes, etc. (e) Trauma; fracture of the base, etc. (f) Hereditary degenerative diseases and malformations of the internal ear. (g) Intoxications, alcohol, nicotine, quinine, salicylic acid group, arsenic. To these may be added gas emboli in caisson disease and ordinary emboli.

**SYMPTOMS.**—The attack usually sets in suddenly with a buzzing noise in the ears and the patient feels as if he were reeling or staggering. He may feel himself to be reeling, or the objects about him may seem to be turning, or the phenomena may be combined. The attack is often so abrupt that the patient falls, though, as a rule, he has time to steady himself by grasping some neighboring object. Consciousness is generally maintained, but may be momentarily lost. Ocular symptoms are usually present. Jerking of the eyeballs, or nystagmus, occurs. The patient becomes pale and nauseated; a clammy sweat breaks out on the face, and vomiting may follow. The duration of the attack varies; it may be very short, but usually the patient has to lie quietly for some time, as any movement of the head brings on another attack. Labyrinthine vertigo is usually paroxysmal, coming on at irregular intervals, sometimes of weeks or months; or several attacks may occur in a day.

The *outlook* is uncertain. While many patients recover completely, in others deafness results and the attacks recur at shorter intervals. In aggra-

vated cases the patient constantly suffers from vertigo, and may even be confined to his bed.

**Acute Vestibulitis.**—In this there is severe vertigo with vomiting and ataxia of a cerebellar form, suggesting bilateral disease of the cerebellum. Permanent deafness on both sides comes on rapidly; the cerebellar features gradually disappear.

**Affections of the External and Middle Ears.**—Irrigation of the meatus may be followed by giddiness or by a severe Ménière syndrome. Wax is one of the commonest causes and the first to be sought for. Removal of a dried fragment pressing against the drum may cure a persistent and distressing vertigo. All forms of *middle ear disease* may cause vertigo, the suppurative as well as the chronic sclerotic. Noises in the ear are usually present as well. The attacks may be of great severity, but apart from gross brain lesions, death is rare. A patient with chronic deafness and tinnitus had severe vertigo in turning in bed on the left side. There was no suggestion of central lesion. Death occurred in one of the attacks.

**(2) Vertigo in Intracranial Tumors.**—The symptom is variable: the largest growths may exist in any region without it—a very small one in a special locality may cause severe attacks. The vestibular fibres may be directly involved in any part of their course or indirectly compressed. Direct involvement is seen in tumors of the cerebello-pontine angle, affecting the eighth and usually the seventh nerves, in tumors of the cerebellum, and in aneurism.

Vertigo is rarely a focal symptom as it may follow indirect pressure from tumors of the cerebrum.

**(3) Ocular Vertigo.**—The association of giddiness with ocular defects has long been recognized, and the newly studied ocular reflexes of vestibular nerve origin now play an important rôle in diagnosis. Nystagmus, double vision, and paralysis of accommodation may be ocular associations of vertigo. The central connections of the nuclei of the "space nerve" with those of the ocular muscles are very close. Errors of refraction may cause an irritation and instability of the space nerve centres leading to severe vertigo.

**(4) Cardio-vascular Vertigo.**—Vertigo is a common feature in neuro-circulatory asthenia. In cardiac insufficiency giddiness is a frequent complaint particularly with aortic disease. The loss of consciousness in Stokes-Adams' disease may be preceded by distressing symptoms of vertigo. One of the commonest forms is seen in hypertension with arterio-sclerosis, very often with tinnitus. It may be slight and noticed only in the morning or on getting up suddenly. In other instances it is one of the most distressing features of progressive sclerosis of the cerebral arteries. Vertigo may precede or accompany the attacks of transient paralysis with aphasia; and with a persistent headache and hypertension it may precede an apoplexy. Low blood pressure is also a frequent cause.

**(5) Toxic vertigo** is described as due to alcohol, tobacco and quinine, to the toxins of the specific fevers, and to focal infection. The essential process is a neuritis of the eighth nerve, or a chronic degenerative change, involving cochlea and labyrinth. A high-pitched tinnitus, with progressive deafness, and transient attacks of vertigo, sometimes of the Ménière type, are the usual symptoms. True toxic neuritis of the vestibular nerve is very

rare. Gastric, renal and various types of functional vertigo have diminished progressively in importance since the studies of Bárány.

**DIAGNOSIS.**—The nervous, anaemic and cardio-vascular groups rarely offer any difficulty but the diagnosis from minor epilepsy is not so easy, particularly in the types without spasm. Tinnitus may be present, but it is rare to have actual loss of consciousness in aural vertigo, in which, also, the actual giddiness is more persistent. The simpler Bárány tests may be applied; the more complicated ones call for the help of the specialist. A full consideration will be found in Barker's *Clinical Diagnosis*, vol. iii. The vestibular reflexes are as important in some cases as those of the iris.

**TREATMENT.**—Bromide of potassium, in 20 grains (1.3 gm.) doses three times a day, is sometimes beneficial. If there is a history of syphilis the iodides should be administered. The salicylates should always be tried in the Ménière syndrome. In cases in which there is hypertension nitroglycerin may be given, at first in very small doses, but increasing gradually. It is not specially valuable in Ménière's disease, but in the giddiness in middle age associated with arterio-sclerosis it sometimes acts very satisfactorily. Correction of errors of refraction is sometimes followed by prompt relief of vertigo.

**Endemic Paralytic Vertigo.**—In parts of Switzerland and France there is a remarkable form of vertigo described by Gerlier, which is characterized by attacks of parietic weakness of the extremities, falling of the eyelids, remarkable depression, but with retention of consciousness. It occurs also in northern Japan, where Miura says it develops paroxysmally among the farm laborers of both sexes and all ages. It is known there as *kubisagari*.

#### GLOSSO-PHARYNGEAL NERVE

The ninth nerve contains both motor and sensory fibres and is also a nerve of the special sense of taste to the tongue. It supplies, by its motor branches, the stylo-pharyngeus and the middle constrictor of the pharynx. The sensory fibres are distributed to the upper part of the pharynx.

**Symptoms.**—Of nuclear disturbance we know very little. The pharyngeal symptoms of bulbar paralysis are probably associated with involvement of the nuclei of this nerve. Lesion of the nerve trunk itself is rare, but it may be compressed by tumors or involved in meningitis. Disturbance of the sense of taste may result from loss of function of this nerve, in which case it is chiefly in the posterior third of the tongue. In neuralgia of the nerve there is pain in the throat about the tonsil and anterior pillars which radiates to the ear and upper part of the neck.

The general disturbances of the sense of *taste* may be briefly mentioned. Loss of the sense of taste—*ageusia*—may be caused by disturbance of the peripheral end organs, as in affections of the mucosa of the tongue. This is common in fever or dyspepsia, in which conditions, as the saying is, everything tastes alike. Strong irritants, such as pepper, tobacco, or vinegar, may dull or diminish the sense of taste. Complete loss may be due to involvement of the nerves either in their course or in the centres. Perversion of the sense of taste—*parageusia*—is rarely found, except as an hysterical manifestation and in the insane. Increased sensitiveness is still more rare. There are

occasional subjective sensations of taste, occurring as an aura in epilepsy or as part of the hallucinations in the insane.

#### PNEUMOGASTRIC (VAGUS) NERVE

The tenth nerve has an important and extensive distribution, supplying the pharynx, larynx, lungs, heart, œsophagus, and stomach. The nerve may be involved at its nucleus along with the spinal accessory and the hypoglossal, forming what is known as bulbar paralysis. It may be compressed by tumors or aneurism, or in the exudation of meningitis, simple or syphilitic. In its course in the neck the trunk may be involved by tumors or in wounds. It has been tied in ligature of the carotid, and has been cut in the removal of deep-seated tumors. The trunk may be attacked by neuritis.

The affections of the vagus are best considered in connection with the distribution of the separate nerves.

**Pharyngeal Branches.**—With the glosso-pharyngeal the branches from the vagus form the pharyngeal plexus, from which the muscles and mucosa of the pharynx are supplied. In *paralysis* due to involvement of this either in the nuclei, as in bulbar paralysis, or in the course of the nerve, as in diphtheritic neuritis, there is difficulty in swallowing and the food is not passed on into the œsophagus. If the nerve on one side only is involved the deglutition is not much impaired. In these cases the particles of food frequently pass into the larynx, and, when the soft palate is involved, into the posterior nares.

**SPASM** of the pharynx is always a functional disorder, usually occurring in hysterical and nervous people. Gowers mentioned a case of a gentleman who could not eat unless alone, on account of the inability to swallow in the presence of others from spasm of the pharynx. This spasm is a well marked feature in hydrophobia, and occurs also in pseudo-hydrophobia.

**Laryngeal Branches.**—The superior laryngeal nerve supplies the mucous membrane of the larynx above the cords and the crico-thyroid muscle. The inferior or recurrent laryngeal curves around the arch of the aorta on the left side and the subclavian artery on the right passes along the trachea and supplies the mucosa below the cords and all the muscles of the larynx except the crico-thyroid and the epiglottidean. Experiments have shown that these motor nerves of the pneumogastric are all derived from the spinal accessory. The remarkable course of the recurrent laryngeal nerves renders them liable to pressure within the thorax, particularly by aneurism. The following are the most important forms of paralysis:

(a) **BILATERAL PARALYSIS OF THE ABDUCTORS.**—In this condition the posterior crico-arytenoids are involved and the glottis is not opened during inspiration. The cords may be close together in the position of phonation, and during inspiration may be brought even nearer together by the pressure of air, so that there is only a narrow chink through which the air whistles with a noisy stridor. This dangerous form of laryngeal paralysis occurs occasionally as a result of cold, or may follow a laryngeal catarrh. The posterior muscles have been found degenerated when the others were healthy. The condition may be produced by pressure upon both vagi, or upon both recurrent nerves. As a central affection it occurs in tabes and bulbar paralysis, but

may be seen also in hysteria. The characteristic symptoms are inspiratory stridor with unimpaired phonation. Possibly, as Gowers suggested, many cases of so-called hysterical spasm of the glottis are in reality abductor paralysis.

(b) **UNILATERAL ABDUCTOR PARALYSIS.**—This frequently results from the pressure of tumors or involvement of one recurrent nerve. Aneurism is the most common cause, though on the right side the nerve may be involved in thickening of the pleura. The left nerve may be involved in mitral stenosis. The symptoms are hoarseness or roughness of the voice, as is so common in aneurism. Dyspnoea is not often present. The cord on the affected side does not move in inspiration. Subsequently the adductors may become involved, in which case phonation is still more impaired.

(c) **ADDUCTOR PARALYSIS.**—This results from involvement of the lateral crico-arytenoid and the arytenoid muscle itself. It is common in hysteria, particularly of women, and causes the hysterical aphonia, which may come on suddenly. It may result from catarrh of the larynx or from overuse of the voice. In laryngoscopic examination it is seen, on attempting phonation, that there is no power to bring the cords together.

(d) **SPASM OF THE MUSCLES OF THE LARYNX.**—In this the adductor muscles are involved. It is not uncommon in children, and has been referred to as laryngismus stridulus. Paroxysmal attacks of laryngeal spasm are rare in the adult, but cases are described in which the patient, usually a young girl, wakes at night in an attack of intense dyspnoea, which may persist long enough to produce cyanosis. Living states that they may replace attacks of migraine. They occur in a characteristic form in tabes, the so-called laryngeal crises. There is a spastic aphonia, in which, when the patient attempts to speak, phonation is completely prevented by a spasm.

Disturbance of the sensory nerves of the larynx is rare.

(e) **ANESTHESIA** may occur in bulbar paralysis and in diphtheritic neuritis—a serious condition, as portions of food may enter the trachea. It is usually associated with dysphagia and is sometimes present in hysteria. Hyperaesthesia of the larynx is rare.

**Cardiac Branches.**—The cardiac plexus is formed by the union of branches of the vagi and of the sympathetic nerves. The vagus fibres subserve motor, sensory and probably trophic functions.

**MOTOR.**—The fibres which inhibit, control, and regulate the cardiac action pass in the vagi. Irritation may produce slowing of the action. Czermak could slow or even arrest the heart's action for a few beats by pressing a small tumor in his neck against one pneumogastric nerve, and it is said that the same can be produced by forcible bilateral pressure on the carotid canal. There are instances in which persons appear to have had voluntary control over the action of the heart. Cheyne mentions the case of Colonel Townshend, "who could die or expire when he pleased, and yet by an effort or somehow come to life again, which it seems he had sometimes tried before he had sent for us." Retardation of the heart's action has also followed accidental ligature of one vagus. Irritation of the nuclei may also be accompanied with a neurosis of this nerve. On the other hand, when there is complete paralysis of the vagi, the inhibitory action may be abolished and the acceleratory influences have full sway. The heart's action is then greatly

increased. This is seen in some instances of diphtheritic neuritis and in involvement of the nerve by tumors, or its accidental removal or ligature. Complete loss of function of one vagus, however, may not be followed by any symptoms.

**SENSORY symptoms** on the part of the cardiac branches are very varied. Normally, the heart's action proceeds regularly without the participation of consciousness, but the unpleasant feelings and sensations of palpitation and pain are conveyed to the brain through this nerve. How far the fibres of the pneumogastric are involved in angina it is impossible to say.

**Pulmonary Branches.**—We know very little of the pulmonary branches of the vagi. The motor fibres are stated to control the action of the bronchial muscles. The various alterations in the respiratory rhythm are probably due more to changes in the centre than in the nerves themselves.

**Gastric and Œsophageal Branches.**—The muscular movements of these parts are presided over by the vagi and vomiting is induced through them, usually reflexly, but also by direct irritation, as in meningitis. Spasm of the œsophagus generally occurs with other nervous phenomena. Pain may be due to cramp of the stomach or to sensory disturbance of this nerve, due to irritation of the peripheral ends, or a neuralgia of the terminal fibres. Some forms of nervous dyspepsia probably depend upon disturbed function of this nerve. The severe gastric crises which occur in tabes are due to central irritation of the nuclei. Vagotonia is an important element in many disorders of the digestive tract.

#### SPINAL ACCESSORY NERVE

**Paralysis.**—The smaller or internal part of this nerve joins the vagus and is distributed through it to the laryngeal muscles. The larger external part is distributed to the sterno-mastoid and trapezius muscles.

The nuclei of the nerve, particularly of the accessory part, may be involved in bulbar paralysis. The nuclei of the external portion, situated in the cervical cord, may be attacked in progressive degeneration of the motor nuclei of the cord. The nerve may be involved in the exudation of meningitis, or be compressed by tumors, or in caries. The *symptoms* of paralysis of the accessory portion which joins the vagus have already been given in the account of the palsy of the laryngeal branches of the pneumogastric. Disease or compression of the external portion is followed by paralysis of the sterno-mastoid and of the trapezius on the same side. In paralysis of one sterno-mastoid the patient rotates the head with difficulty to the opposite side, but there is no torticollis, though in some cases the head is held obliquely. As the trapezius is supplied in part from the cervical nerves, it is not completely paralyzed, but the portion which passes from the occipital bone to the acromion is functionless. The paralysis of the muscle is well seen when the patient draws a deep breath or shrugs the shoulders. The middle portion of the trapezius is also weakened, the shoulder droops a little, and the angle of the scapula is rotated inward by the action of the rhomboids and the levator anguli scapulæ. Elevation of the arm is impaired, for the trapezius does not fix the scapula as a point from which the deltoid can work.

In progressive muscular atrophy we sometimes see bilateral paralysis of these muscles. Thus, if the sterno-mastoids are affected, the head tends to fall back; when the trapezii are involved, it falls forward, a characteristic attitude of the head in many cases of progressive muscular atrophy. Gowers suggested that lesions of the accessory in difficult labor may account for those cases in which during the first year of life the child has great difficulty in holding up the head. In children this drooping of the head is an important symptom in cervical meningitis, the result of caries.

The TREATMENT of the condition depends much upon the cause. In the central nuclear atrophy but little can be done. In paralysis from pressure the symptoms may gradually be relieved. The paralyzed muscles should be stimulated by electricity and massage.

**Accessory Spasm** (*Torticollis; Wryneck*).—The forms of spasm affecting the cervical muscles are best considered here, as the muscles supplied by the accessory are chiefly, though not solely, responsible for the condition.

(a) CONGENITAL TORTICOLLIS.—This condition, also known as *fixed* torticollis, depends upon the shortening and atrophy of the sterno-mastoid on one side. It occurs in children and may not be noticed for several years on account of the shortness of the neck, the parents often alleging that it has only recently come on. It affects the right side almost exclusively. A remarkable circumstance in connection with it is the existence of facial asymmetry noted by Wilks, which appears to be an essential part of this congenital form. In congenital wryneck the sterno-mastoid is shortened, hard and firm, and in a condition of more or less advanced atrophy. This must be distinguished from the local thickening in the sterno-mastoid due to rupture, which may occur at birth and produce an induration. Although the sterno-mastoid is almost always affected, there are rare cases in which the fibrous atrophy affects the trapezius. This form of wryneck is readily relieved by tenotomy, but Gölding-Bird states that the facial asymmetry persists, or may become more evident. With reference to the pathology, Gölding-Bird concludes that the facial asymmetry and the torticollis are integral parts of one affection which has a central origin, and is the counterpart in the head and neck of infantile paralysis with talipes in the foot.

(b) SPASMODIC WRYNECK.—Two varieties of this spasm occur, the tonic and the clonic, which may alternate in the same case; or, as is most common, they are separate and remain so from the outset. The disease is most frequent in adults and, according to Gowers, more common in females. In America it is certainly more frequent in males. In females it may be an hysterical manifestation. There may be a marked neurotic family history, but it is usually impossible to fix upon any definite etiological factor. Some cases have followed cold; others a blow. Brissaud described what he calls mental torticollis. It is usually met with in neurasthenic patients and in elderly persons, and consists of a clonic spasm of the rotators of the head.

The *symptoms* are well defined. In the *tonic* form the contracted sterno-mastoid draws the occiput toward the shoulder of the affected side; the chin is raised, and the face rotated to the other shoulder. The sterno-mastoid may be affected alone or in association with the trapezius. When the latter is implicated the head is depressed still more toward the same side. In long-

standing cases these muscles are prominent and very rigid. There may be some curvature of the spine, the convexity of which is toward the sound side. The cases in which the spasm is *clonic* are much more distressing and serious. The spasm is rarely limited to a single muscle. The sterno-mastoid is almost always involved and rotates the head so as to approximate the mastoid process to the inner end of the clavicle, turning the face to the opposite side and raising the chin. When with this the trapezius is affected, the depression of the head toward the same side is more marked. The head is drawn somewhat backward; the shoulder, too, is raised by its action. The splenius may be associated with the sterno-mastoid. Its action is to incline the head and rotate it slightly toward the same side. Other muscles may be involved, such as the scalenus and platysma myoides; and in rare cases the head may be rotated by the deep cervical muscles. There are cases in which the spasm is bilateral, causing a backward movement—retro-colic spasm. This may be tonic or clonic; in extreme cases the face is horizontal and looks upward.

These clonic contractions may come on without warning, or be preceded by irregular pains or stiffness of the neck. The jerking movements recur every few moments, and it is impossible to keep the head still for more than a minute or two. In time the muscles undergo hypertrophy and may be distinctly larger on one side than the other. In some cases the pain is considerable; in others there is simply a feeling of fatigue. The spasms cease during sleep. Emotion, excitement, and fatigue increase them. The spasm may extend from the neck muscles and involve those of the face or arms.

The disease varies much; cases occasionally get well, but the majority persist, and, even if temporarily relieved, the disease frequently recurs. The affection is usually regarded as a functional neurosis, but it is possibly due to disturbance of the cortical centres presiding over the muscles.

*Treatment.*—Temporary relief is sometimes obtained; a permanent cure is exceptional. Psycho-therapeutic treatment has been successful in some cases. In mild cases education in the voluntary relaxation of the affected muscles is useful. Various drugs have been used, but rarely with benefit. Occasionally, large doses of bromide lessen the intensity of the spasms. Morphia has been successful in some cases, but there is great danger of establishing a habit. Galvanism may be tried. Counter-irritation is probably useless. Fixation of the head mechanically can rarely be borne by the patient. These obstinate cases come ultimately to the surgeon, and the operations of stretching, division, and excision of the accessory nerve and division of the muscles have been tried. Temporary relief may follow and is in proportion to the extent of the operation, but, as a rule, the condition returns. Risien Russell thinks that resection of the posterior branches of the upper cervical nerves is most likely to give relief.

(c) The **NODDING SPASM** of children may here be mentioned as involving chiefly the muscles innervated by the accessory nerve. It may be a simple trick, a form of habit spasm, or a phenomenon of epilepsy (*E. nultans*), in which case it is associated with transient loss of consciousness. A similar nodding spasm may occur in older children. In women it sometimes occurs as an hysterical manifestation, commonly as part of the so-called salaam convulsion.

## HYPOGLOSSAL NERVE

This is the motor nerve of the tongue and for most of the muscles attached to the hyoid bone. Its cortical centre is probably the lower part of the anterior central gyrus.

**Paralysis.**—(a) CORTICAL LESION.—The tongue is often involved in hemiplegia, and the paralysis may result from a lesion of the cortex itself, or of the fibres as they pass to the medulla. It does not occur alone and is considered with hemiplegia. There is this difference, however, between the cortical and other forms, that the muscles on both sides of the tongue may be more or less affected but do not waste, nor are their electrical reactions disturbed.

(b) NUCLEAR and INFRA-NUCLEAR lesions result from slow progressive degeneration, as in bulbar paralysis or tabes; occasionally there is acute softening from obstruction of the vessels. The nuclei of both nerves are usually affected together, but may be attacked separately. Trauma, syphilis and lead poisoning are causes. The fibres may be damaged by a tumor, and at the base by meningitis; or the nerve may be involved in the condylar foramen by disease of the skull. It may be involved in its course in a scar, as in Birkett's case, or compressed by a tumor in the parotid region. As a result, there is loss of function in the nerve fibres and the tongue undergoes atrophy on the affected side. It is protruded toward the paralyzed side and may show fibrillary twitching.

The *symptoms* of involvement of one hypoglossal, either at its centre or in its course, are those of unilateral paralysis and atrophy of the tongue. When protruded, it is pushed toward the affected side, and there are fibrillary twitchings. The atrophy is usually marked and the mucous membrane on the affected side is thrown into folds. Articulation is not much impaired in the unilateral affection. When the disease is bilateral, the tongue lies almost motionless in the floor of the mouth; it is atrophied, and can not be protruded. Speech and mastication are extremely difficult and deglutition may be impaired. If the seat of the disease is above the nuclei, there may be little or no wasting. The condition is seen in progressive bulbar paralysis and occasionally in progressive muscular atrophy.

The *diagnosis* is readily made and the situation of the lesion can usually be determined, since when supra-nuclear there is associated hemiplegia and no wasting of the muscles of the tongue. Nuclear disease is only occasionally unilateral; most commonly bilateral and part of a bulbar paralysis. The fibres of the hypoglossal may be involved within the medulla after leaving their nuclei. In such a case there may be paralysis of the tongue on one side and paralysis of the limbs on the opposite side, and the tongue, when protruded, is pushed toward the sound side. The *treatment* is that of the causal condition.

**Spasm.**—This rare affection may be unilateral or bilateral. It is most frequently a part of some other convulsive disorder, such as epilepsy, chorea, or spasm of the facial muscles. In some cases of stuttering, spasm of the tongue precedes the explosive utterance of the words. It may occur in hysteria, and is said to follow reflex irritation in the fifth nerve. The most remarkable cases are those of paroxysmal clonic spasm, in which the tongue

is rapidly thrust in and out, as many as forty or fifty times a minute. The prognosis is usually good.

#### COMBINED PARALYSIS OF THE LAST THREE AND FOUR CRANIAL NERVES

The war experience has widened our knowledge of these cases. There may be: (a) *Avelli's syndrome*, palato-laryngeal paralysis from involvement of the ninth and eleventh. With this there may be involvement of the tenth with paralysis of the superior constrictor of the pharynx. When the outer fibres of the spinal accessory are involved, the sterno-cleido-mastoid may be paralyzed on the same side (*Schmidt's syndrome*). (b) *Hughlings-Jackson's syndrome*. Involvement of the ix, x, xi, and xii—disturbance of taste and paralysis of the superior constrictor of the pharynx (ix and x); hemi-anæsthesia of the palate and pharynx, sometimes with cough and dyspnoea and salivation which may be profuse (x and xi); hemi-paralysis of the larynx (xi) with hemi-paralysis of the tongue (xii). In wounds of the retro-parotidian space or after a parotid bubo, in addition to the hypoglossal, the sympathetic nerves with fibres of the ix, x, and xi may be involved, causing exophthalmos, myosis, and sweating, with the combined paralyse known as Villaret's syndrome. These combined paralyse may be nuclear caused by gummatous or tuberculous meningitis, by tumor or by injury. In the war cases the lesions have often been more extensive, and symptoms of involvement of the vagus have been more common than in the ordinary instances from tumor or meningitis.

### IV. DISEASES OF THE SPINAL NERVES

#### CERVICAL PLEXUS

**Occipito-cervical Neuralgia.**—This involves the nerve territory supplied by the occipitalis major and minor, and the auricularis magnus nerves. The pains are chiefly in the back of the head and neck and in the ear. The condition may follow cold and is sometimes associated with stiffness of the neck or torticollis. Unless disease of the bones exists with it or it is due to pressure of tumors, the outlook is usually good. There are tender points midway between the mastoid process and the spine and just above the parietal eminence, and between the sterno-mastoid and the trapezius. The affection may be due to direct pressure in carrying heavy weights.

**Affections of the Phrenic Nerve.**—Paralysis may follow a lesion in the anterior horns at the level of the third and fourth cervical nerves, or may be due to compression of the nerve by tumors or aneurism. More rarely paralysis results from neuritis, alcoholic, diphtheritic or saturnine.

When the *diaphragm* is paralyzed respiration is carried on by the intercostal and accessory muscles. When the patient is quiet and at rest little may be noticed, but the abdomen retracts in inspiration and is forced out in expiration. On exertion or even on attempting to move there may be dyspnoea. If the paralysis sets in suddenly there may be dyspnoea and lividity, which is usually temporary (W. Pasteur). Pneumonia or bronchitis seriously aggra-

vates the condition. Difficulty in coughing, owing to the impossibility of drawing a full breath, adds greatly to the danger.

When the phrenic nerve is paralyzed on one side the paralysis may be scarcely noticeable, but inspection and the fluoroscope show that the descent of the diaphragm is much less on the affected side.

The *diagnosis* of paralysis is not always easy, particularly in women, who habitually use this muscle less than men, and in whom the diaphragmatic breathing is less conspicuous. Immobility of the diaphragm is not uncommon, particularly in diaphragmatic pleurisy, in large effusions, and in extensive emphysema. The muscle itself may be degenerated.

Owing to the lessened action of the diaphragm, there is a tendency to stasis at the bases of the lungs, and there may be impaired resonance and signs of oedema. As a rule, however, the paralysis is not confined to this muscle, but is part of a general neuritis or a polio-myelitis, and there are other symptoms of value in determining its presence. The outlook is usually serious. The treatment is that of the neuritis or polio-myelitis. Artificial respiration should be carried on if necessary.

**Hiccough.**—Here may be considered this remarkable symptom, caused by intermittent, sudden contraction of the diaphragm. The mechanism, however, is complex, and while the afferent impressions to the respiratory centre may be peripheral or central the efferent are distributed through the phrenic nerve to the diaphragm, causing the intermittent spasm, and through the laryngeal branches of the vagus to the glottis, causing sudden closure as the air is rapidly inspired. There are various groups:

(a) **INFLAMMATORY**, seen particularly in affections of the abdominal viscera, gastritis, peritonitis, hernia, internal strangulation, appendicitis, suppurative pancreatitis, and in severe forms of typhoid fever.

(b) **IRRITATIVE**, as in the direct stimulation of the diaphragm when very hot substances are swallowed, in disease of the œsophagus near the diaphragm, and in many conditions of gastric and intestinal disorder, more particularly those associated with flatus.

(c) **TOXIC**.—In these cases there is usually some general disease, as gout, diabetes, or chronic nephritis. Hiccough may be very obstinate in the later stages of chronic nephritis.

(d) Cases in which the primary cause is in the *nervous system*; hysteria, epilepsy, shock, or cerebral tumors. It may be persistent in epidemic encephalitis.

The **TREATMENT** is often very unsatisfactory. Sometimes in the milder forms a sudden reflex irritation will check it at once. A pinch of snuff may be effective. Readers of Plato's Symposium will remember that the physician Eryximachus recommended to Aristophanes, who had hiccough from eating too much, either to hold his breath (which for trivial forms of hiccough is very satisfactory) or to gargle with a little water; but if it still continued, "tickle your nose with something and sneeze; and if you sneeze once or twice even the most violent hiccough is sure to go." The attack must have been of some severity, as it is stated subsequently that the hiccough did not disappear until Aristophanes had resorted to the sneezing.

Ice, a teaspoonful of salt and lemon juice, or salt and vinegar, or a teaspoonful of raw spirits may be tried. When the hiccough is due to gastric

irritation, lavage is sometimes promptly curative. Alkali should be given freely. A hypodermic injection of gr.  $\frac{1}{8}$  (0.008 gm.) of apomorphia may give prompt relief. In obstinate cases the various antispasmodics have been used in succession. Pilocarpine has been recommended. The ether spray on the epigastrium may be effective. Hypodermics of morphia, inhalations of chloroform, the use of nitrite of amyl and of nitroglycerin have been beneficial in some cases. Benzyl benzoate in doses of 30 drops of a 20 per cent. solution sometimes is effectual. Galvanism over the phrenic nerve, or pressure on the nerves, applied between the heads of the sterno-cleido-mastoid muscles may be used. Strong traction upon the tongue may give immediate relief. Of all measures morphia used freely is the best.

#### BRACHIAL PLEXUS

**Cervical Rib.**—FREQUENCY.—The anomaly is much more common than indicated in the literature. Sometimes bilateral, it may be complete with bony attachment to the second rib; incomplete, forming a short stump of variable length, or—and this is important—there may be a fibrous band-like attachment from a short rib to the first. It is more common on the left side. Symptoms usually appear between the fifteenth and thirtieth years, and their onset is often due to dropping of the shoulder girdle. Some form of exercise or in women the carrying of a child on the arm may be responsible.

The ribs may be visible, one more plainly than the other, and the subclavian artery, lifted up, may pulsate high in the supraclavicular fossa. This abnormal pulsation and the fullness in the fossa may suggest the presence of the extra rib. The throbbing may be marked enough to suggest aneurism. The rib may be felt, often more marked on one side; even the bifid extremity may be palpable, and the artery felt above the rib sometimes appears longer and larger than normal.

**SYMPTOMS.**—Many patients are unaware of the anomaly; the symptoms, which may come on suddenly, may be grouped as follows:

(1) *Local.* (a) Supraclavicular swelling. (b) Pulsation. (c) Palpable tumor and aneurism.

(2) *Neuritic.* (a) Neuralgic pains (supraclavicular, cervical, brachial). (b) Paræsthesia. (c) Local anæsthesia. (d) Sympathetic nerve features.

(3) *Muscular.* (a) Atrophy, in ulnar distribution. (b) *Spasm.* (c) Intermittent claudication.

\*(4) *Vascular.* (a) Vaso-motor changes (ischæmia, hyperæmia, swelling). (b) Local gangrene. (c) Aneurism, (i) spurious, (ii) true. (d) Thrombosis.

Neuralgic pains occur in the cervical region, sometimes passing up the back of the head; more commonly the pain is in the distribution of the eighth cervical and first dorsal nerve, sometimes only a dull pain and aching with numbness and tingling or even anæsthesia. Dissociation of cutaneous sensation, loss of tactile and thermic with retention of pain sense, may be present. The cervical sympathetic may be involved with the usual features. *Muscular atrophy* is usually in the region of distribution of the ulnar nerve. The difference between the two arms may be marked and the interossei wasted, as in progressive muscular atrophy, for which, when bilateral, cases may be mistaken. With pressure on and narrowing of the subclavian, intermittent

claudication is present, characterized by numbness, tingling and swelling, sometimes by redness of the arm and muscular disability on exertion. At rest the arm is normal and comfortable, but on exertion these features occur: spasm, tonic or clonic, in the muscles of the hand is occasionally seen.

**VASO-MOTOR CHANGES.**—Redness with swelling, sometimes cyanosis and mottling, may be present, with changes resembling Raynaud's disease; in a few cases gangrene of the finger tips has followed.

**ANEURISM.**—The subclavian artery may be tilted by the ribs and give a wide area of supraclavicular pulsation. There may be: (1) Slight narrowing from pressure, with feeble pulse on the affected side; (2) manifest enlargement of the vessel, fusiform or uniform; or (3) a definite cylindrical aneurism. In 27 of 525 clinical cases collected by Halsted these local changes were present. The dilatation is distal to the point of constriction made by the rib and the scalenus anticus, which Halsted explains by the abnormal play of the blood in the relatively dead pocket beyond the constriction, and the absence of the normal pulse pressure necessary to maintain the integrity of the arterial wall. The *nervi arteriorum* may be involved.

**THROMBOSIS.**—This may occur in the vessels beyond the point of constriction, in one case involving suddenly the brachial and gradually extending to the axillary and subclavian, with the gradual development of an effective collateral circulation.

The relative distribution of the symptoms as given by Halsted from an exhaustive review of the literature was in 63.3 per cent. nerve symptoms alone, in 29.4 per cent., nervous and vascular symptoms, while 5.3 per cent., have only vascular symptoms.

**DIAGNOSIS.**—This is easy as a rule even without the X-rays. A serious difficulty arises when disease of the cord occurs in the subjects of cervical rib, e. g., syringomyelia and progressive muscular atrophy. In cases of prolonged discomfort or pain with vascular or trophic disturbance in the arm, cervical rib should be considered.

**TREATMENT.**—When accidentally discovered, it is best not to tell the patient. Elevation of the shoulders may give relief. Massage, electricity and other forms of local treatment may be tried. The rib may be removed, but only as a last resort, as the results are not always satisfactory.

**Combined Paralysis.**—The plexus may be involved in the supraclavicular region by compression of the nerve trunks as they leave the spine, or by tumors and other morbid processes in the neck. Below the clavicle lesions are more common and result from injuries following dislocation or fracture, sometimes from neuritis. A cervical rib may lead to a pressure paralysis of the lower cord of the plexus. A not infrequent injury in this region follows falls or blows on the neck, which by lateral flexion of the head and depression of the shoulder seriously stretch the plexus. The entire plexus may be ruptured and the arm totally paralyzed. The rupture may occur anywhere between the vertebræ and the clavicle, and involve all the cords of the plexus, or only the upper ones. The so-called "obstetrical palsy" usually results from the forcible separation of the head and neck from the shoulder during delivery, with tearing of the deep cervical fascia and the nerves, involving the roots from above and downward, so that the injury may vary from a slight lesion of the upper root to complete rupture of the plexus or the tearing of

the roots from the cord. In the complete lesion the arm is flaccid and immobile, does not grow, and there is displacement of the head of the humerus; sensory disturbances are rare. The prognosis is bad; only mild cases recover completely. Suturing the broken cords and planting them in the neighboring roots have been followed by good results, but complete recovery rarely if ever follows. Another common cause of lesion of the brachial plexus is luxation of the head of the humerus, particularly the subcoracoid form.

A primary neuritis of the brachial plexus is rare. More commonly the process is an ascending neuritis from a lesion of a peripheral branch, involving first the radial or ulnar nerves, and spreading upward to the plexus, producing gradually complete loss of power in the arm.

**Lesions of Individual Nerves of the Plexus.**—(a) **LONG THORACIC NERVE.**—*Serratus paralysis* follows injury to this nerve in the neck, usually by direct pressure in carrying loads, and is very common in soldiers. It may be due to a neuritis following an acute infection or exposure. Isolated serratus paralysis is rare. It usually occurs with paralysis of other muscles of the shoulder girdle, as in the myopathies and progressive muscular atrophy. Concomitant trapezius paralysis is the most frequent. In the isolated paralysis there is little or no deformity with the hands hanging by the sides. There are slight abnormal obliquity of the posterior border of the scapula and prominence of the inferior angle, but when, as is common, the middle part of the trapezius is also paralyzed the deformity is marked. The shoulder is lower, the inferior angle of the scapula is displaced inward and upward, and the superior angle projects upward. When the arms are held out in front at right angles to the body the scapula becomes winged and stands out prominently. The arm can not, as a rule be raised above the horizontal. The outlook of the cases due to injury or neuritis is good.

(b) **CIRCUMFLEX NERVE.**—This supplies the deltoid and teres minor and may be involved in injuries, in dislocations, bruising by a crutch, or sometimes by extension from arthritis. Occasionally the paralysis arises from a pressure neuritis during an illness. As a consequence of loss of power in the deltoid, the arm can not be raised. The wasting is usually marked and changes the shape of the shoulder. Sensation may be impaired in the skin over the muscle. The joint may be relaxed and there may be a distinct space between the head of the humerus and the acromion.

(c) **MUSCULO-SPIRAL PARALYSIS; RADIAL PARALYSIS.**—This is one of the most common of peripheral palsies, due to the exposed position of the musculospiral nerve. It is often bruised by a crutch, by injuries of the arm, blows, or fractures. It is frequently injured when a person falls asleep with the arm over the back of a chair, or by pressure of the body upon the arm when a person is sleeping on a bench or on the ground. It may be paralyzed by sudden violent contraction of the triceps. It is sometimes involved in a neuritis from cold, but this is uncommon in comparison with other causes. The paralysis of lead poisoning is the result of involvement of certain branches of this nerve.

A lesion when high up involves the triceps, the brachialis anticus, and the supinator longus, as well as the extensors of the wrist and fingers. In lesions just above the elbow the arm muscles and the supinator longus are spared. The most characteristic feature is the wrist-drop and the inability

to extend the first phalanges of the fingers and thumb. In the pressure palsies the supinators are usually involved, and the movements of supination can not be accomplished. Sensation may be impaired, or there may be marked tingling, but the loss of sensation is rarely so pronounced as that of motion.

The affection is readily recognized but it is sometimes difficult to say upon what it depends. The sleep and pressure palsies are, as a rule, unilateral and involve the supinator longus. The paralysis from lead is bilateral and the supinators are unaffected. Bilateral wrist-drop is a very common symptom in many forms of multiple neuritis, particularly the alcoholic; but the mode of onset and the involvement of the legs and arms make the diagnosis easy. The duration and course of the musculo-spiral paralyses are very variable. The pressure palsies may disappear in a few days. Recovery is the rule, even when the affection lasts for many weeks. The electrical examination is of importance in prognosis, and the rules laid down under paralysis of the facial nerve hold good here. The *treatment* is that of neuritis.

(d) ULNAR NERVE.—The motor branches supply the ulnar half of the deep flexor of the fingers, the muscles of the little finger, the interossei, the adductor and the inner head of the short flexor of the thumb, and the ulnar flexor of the wrist. The sensory branches supply the ulnar side of the hand—two (or one) and a half fingers on the back, and one and a half fingers on the front. Paralysis may result from pressure, usually at the elbow joint, although the nerve is here protected. Possibly the neuritis in the ulnar nerve in some cases of acute illness may be due to this cause. Owing to paralysis of the ulnar flexor of the wrist, the hand moves toward the radial side; adduction of the thumb is impossible; the first phalanges can not be flexed, and the others can not be extended. In long-standing cases the first phalanges are overextended and the others strongly flexed, producing the claw-hand; but this is not so marked as in progressive muscular atrophy. The loss of sensation corresponds to the sensory distribution just mentioned.

(e) MEDIAN NERVE.—This supplies the flexors of the fingers except the ulnar half of the deep flexors, the abductor and the flexors of the thumb, the two radial lumbricales, the pronators, and the radial flexor of the wrist. The sensory fibres supply the radial side of the palm and the front of the thumb, the first two fingers and half the third finger, and the dorsal surfaces of the same three fingers.

This nerve is seldom involved alone. Paralysis results from injury and occasionally from neuritis. The signs are inability to pronate the forearm beyond the mid-position. The wrist can be flexed only toward the ulnar side; the thumb can not be opposed to the tips of fingers. The second phalanges can not be flexed on the first; the distal phalanges of the first and second fingers can not be flexed; but in the third and fourth fingers this action can be performed by the ulnar half of the flexor profundus. The loss of sensation is in the region corresponding to the sensory distribution already mentioned. The wasting of the thumb muscles, which is usually marked in this paralysis, gives to it a characteristic appearance.

**Volkman's Paralysis.**—*Ischæmic* paralysis, as it is called, usually follows the pressure of splints and bandages in children with fracture in the region of the elbow-joint. The changes are thought to be due to arrest of the circu-

lation in the muscles, which are hardened and stiff and the flexors of the forearm are contracted. The hand is claw-like with the metacarpo-phalangeal joints strongly extended and the middle and terminal phalanges strongly flexed. The condition may come on with great rapidity and appears to be a muscular lesion though it is not always possible to exclude pressure on the nerves. The prognosis is good with judicious treatment.

#### LUMBAR AND SACRAL PLEXUSES

**Lumbar Plexus.**—The lumbar plexus is sometimes involved in growths of the lymph glands, in psoas abscess, and in disease of the bones of the vertebræ. The *obturator nerve* is occasionally injured during parturition. When paralyzed the power is lost over the adductors of the thigh and one leg can not be crossed over the other. Outward rotation is also disturbed. The *anterior crural nerve* is sometimes involved in wounds or in dislocation of the hip-joint, less commonly during parturition, and sometimes by disease of the bones and in psoas abscess. The special symptoms of affection of this nerve are paralysis of the extensors of the knee with wasting of the muscles, anæsthesia of the antero-lateral parts of the thigh and of the inner side of the leg to the big toe. This nerve is sometimes involved early in growths about the spine, and there may be pain in its area of distribution. Loss of the power of abducting the thigh results from paralysis of the *gluteal nerve*, which is distributed to the gluteus medius and minimus muscles.

**External Cutaneous Nerve.**—A peculiar form of sensory disturbance, confined to the territory of this nerve, was first described by Bernhardt in 1895, and a few months later by Roth, who gave it the name of *meralgia paræsthetica*. The disease is probably due to a neuritis which seems to originate in the nerve where it passes under Poupart's ligament, just internal to the anterior superior iliac spine. The nerve is usually tender on pressure at this point. The disease is more common in men. Musser and Sailer in 1900 collected 99 cases, of which 75 were in men. A large number of the cases are attributable to direct trauma or to simple pressure on the nerve in the aponeurotic canal through which it passes. Pregnancy is among the more common causes in women.

The *sensory* disturbances consist of various forms of paræsthesia located over the outer side of the thigh, sometimes with diminished sensation or hyperæsthesia. The symptoms may persist for years with such discomfort, exaggerated by walking or the touch of the clothing, that patients may be greatly incapacitated. Excision of the nerve as it passes under Poupart's ligament has given good results.

**Sacral Plexus.**—The sacral plexus is frequently involved in tumors and inflammations within the pelvis and may be injured during parturition. Neuritis is common, usually an extension from the sciatic nerve.

Goldthwaite calls attention to the fact that the lumbo-sacral articulation varies very greatly in its stability, and displacement of the bones may result with separation of the posterior portion of the intervertebral disc. The cauda equina, or the nerve roots, may be compressed. With displacement on one side the spine is rotated and the articular process of the fifth is drawn into the spinal canal, with such narrowing that paraplegia may result, and he

reports a remarkable case in which paralysis came on during the application of a plaster jacket. Weakness of the joints or displacements may cause irritation of the nerves inside and outside the canal with resulting sciatica.

Of the branches, the *sciatic nerve*, when injured at or near the notch, causes paralysis of the flexors of the legs and the muscles below the knee, but injury below the middle of the thigh involves only the latter muscles. There is also anæsthesia of the outer half of the leg, the sole, and the greater portion of the dorsum of the foot. Wasting of the muscles and trophic disturbances may follow. In paralysis of one sciatic the leg is fixed at the knee by the action of the quadriceps extensor and the patient is able to walk.

Paralysis of the *small sciatic nerve* is rarely seen. The *gluteus maximus* is involved and there may be difficulty in rising from a seat. There is a strip of anæsthesia along the back of the middle third of the thigh.

*External Popliteal Nerve.*—Paralysis involves the peronæi, the long extensor of the toes, *tibialis anticus*, and the *extensor brevis digitorum*. The ankle can not be flexed, resulting in a condition known as foot-drop, and as the toes can not be raised the whole leg must be lifted, producing the characteristic *steppage* gait seen in so many forms of peripheral neuritis. In long-standing cases the foot is permanently extended and there is wasting of the anterior tibial and peroneal muscles. The loss of sensation is in the outer half of the front of the leg and on the dorsum of the foot.

*Internal Popliteal Nerve.*—When paralyzed, plantar flexion of the foot and flexion of the toes are impossible. The foot can not be adducted, nor can the patient rise on tiptoe. In long-standing cases *talipes calcaneus* follows and the toes assume a claw-like position from secondary contracture, due to over-extension of the proximal and flexion of the second and third phalanges.

## SCIATICA

**Definition.**—The term *sciatica* is applied to any painful condition referred to the sciatic nerve. It may be defined as an interstitial inflammation of the sciatic nerve, a *neuro-fibrositis*, causing severe pain in the branches of distribution and, if long continued, atrophy of the muscles. Only rarely does marked motor paralysis or sensory loss result.

**Etiology.**—Primary neuritis of this nerve is very rare and is seen chiefly in diabetes and gout. In the vast majority the condition is *secondary* to a process elsewhere which affects the component cords or the trunk itself. Among the causal factors are: (1) *Arthritis* which may be of the lower spine, lumbosacral, sacro-iliac or hip joints. In this case the arthritic lesion is often due to a focus of infection. (2) Anatomical *anomalies*, as an unusually long transverse process of the fifth lumbar vertebra. (3) Disease of the bones of the lower spine or pelvis, e. g., tuberculosis. (4) *Strain*, which may be acute or chronic, especially of the sacro-iliac joint. Exposure to cold after heavy muscular exertion is said to be a cause. In trench warfare the men were not as subject to sciatica as the officers. (5) *Pelvic* conditions, such as a solid ovarian or fibroid tumor in women and prostatic disease in men. Constipation and the pressure of the fetal head in labor are occasional causes. (6) *Syphilis* is responsible in a few cases. (7) It may be due to a focus of infection, which may cause fibrositis. (8) Among rare causes are an abnormal

network of veins on the trunk and anomalies, such as the piriform muscle passing through the nerve.

**Symptoms.**—Pain is the most constant and troublesome symptom. The onset may be severe, with slight pyrexia, but, as a rule, it is gradual, and for a time there is only slight pain in the back of the thigh, particularly in certain positions or after exertion. Soon the pain becomes more intense, and, instead of being limited to the upper portion of the nerve, extends down the nerve. The patient can often point out the most sensitive spots, usually at the notch or in the middle of the thigh; and on pressure these are exquisitely painful. The pain may occur particularly in the distribution of one of the branches, as the external popliteal. The pain is gnawing or burning, and is usually constant, but in some instances is paroxysmal, and often worse at night. On walking it may be very great; the knee is bent and the patient treads on the toes, so as to relieve the tension on the nerve. In protracted cases there may be wasting of the muscles, but the reaction of degeneration can seldom be obtained. In chronic cases cramp and fibrillary contractions may occur. Herpes may develop but this is unusual. The patient assumes the position in which there is least tension on the nerve and any position or movement which stretches the nerve increases the pain. The knee-jerk is usually increased; the ankle-jerk is decreased or lost. The ankle-jerk may be absent for a long period afterwards. In rare instances the neuritis ascends and involves the spinal cord.

**Duration and Course.**—The duration and course are extremely variable and depend greatly on the cause. As a rule, it is an obstinate affection, lasting for months, or even, with slight remissions, for years. Relapses are not uncommon, and the disease may be relieved in one nerve only to appear in the other. In the severer forms the patient is bedridden, and such cases prove among the most distressing and trying which the physician is called upon to treat.

**Diagnosis.**—It is important, in the first place, to determine whether the disease is primary, or secondary to some affection elsewhere. The diagnosis should determine the cause; lesions of the lower spine and sacro-iliac joints should be searched for especially. A careful rectal examination should be made, and, in women, pelvic tumor should be excluded. "Lumbago" may be confounded or associated with it. Affections of the hip-joint are easily distinguished by the absence of tenderness in the course of the nerve and the pain on movement of the hip-joint or on pressure in the region of the trochanter. Pressure on the nerve trunks of the cauda equina, as a rule, causes bilateral pain and disturbances of sensation, and, as double sciatica is rare, these always suggest lesion of the nerve roots. Pressure on the nerve roots by neoplasm must always be excluded. Between the lightning pains of tabes and sciatica the differences are usually well defined. In a certain number of cases the condition is a fibrositis. There is no tenderness along the course of the sciatic nerve, but there is pain in the gluteal region, with disability and Lasègue's sign, i. e., inability to extend the leg completely when the thigh is flexed on the abdomen.

**Treatment.**—If the cause can be determined, treatment should be directed to correcting this as soon as possible. So many are due to bone conditions which themselves are secondary to disease elsewhere (such as foci of infec-

tion) that a very complete study is necessary. The removal of an infected tooth may cause a rapid improvement. In cases with diabetes or gout the usual treatment for these should be carried out. In all cases certain palliative measures are indicated and may be the only ones available in some cases. The most important is *rest* which should be absolute and in the position which gives the most relief. Fixation of the leg by a splint may be of aid. The patient should not be allowed up for any purpose. The application of heat in some form is helpful. An electric pad, a hot water bag or the cautery may be used. Counter-irritation, especially by blisters, sometimes gives relief. Acupuncture is worth a trial in obstinate cases. Injections into the nerve have been frequently used and various solutions have been employed, e. g., sterile water or novocaine. Exposure of the nerve and incision of the sheath are indicated in severe cases. Electricity may give temporary relief but is often disappointing. X-ray exposures over the roots along the fourth and fifth lumbar and first four sacral vertebræ may be helpful. In some cases time, usually months, seems necessary.

As to drugs, sedatives are usually necessary, the simple ones being preferred, and morphia avoided if possible. The coal-tar products and salicylates in full doses are worth a trial and often give relief when combined with codeine. The use of sedative suppositories is often especially helpful. If there is any suspicion of syphilis, active treatment should be given.

## V. HERPES ZOSTER

### (*Acute Posterior Ganglionitis*)

**Definition.**—An acute disease with localization in the cerebral ganglia and in the ganglia of the posterior nerve roots, associated with a vesicular inflammation of the skin of the corresponding cutaneous areas.

**Distribution.**—Herpes most frequently occurs in the region of the dorsal roots and extends in the form of a half girdle, on which account the names "*zona*" and "*zoster*" have been given. The trigeminal region may be involved, particularly the first branch.

**Etiology.**—It occurs with the acute infections, particularly pneumonia, malaria and cerebro-spinal fever. A streptococcus infection is suggested in some cases. Epidemics have been described. In some cases, especially those in the lower part of the body, syphilis co-exists. Even in non-syphilitic cases, the spinal fluid may show increase in the cells but the globulin is rarely much increased. Herpes zoster may occur with traumatic paraplegia or injury to the ganglia (fracture) or tumors may be responsible. It has followed the administration of arsenic. A curious association of occurrence with chicken-pox has been noted.

**Pathology.**—Bärensprung first showed that there was involvement of the spinal ganglia. The disease is an acute hæmorrhagic inflammation of the ganglia of the posterior nerve roots and of the homologous cranial ganglia (Head and Campbell). There are inflammatory foci, hæmorrhage in and destruction of certain of the ganglion cells leading to degeneration of the axicylinders. In herpes facialis accompanying pneumonia W. T. Howard has

shown that similar lesions are demonstrable in the Gasserian ganglion, and Hunt found the same changes in the otic ganglion in herpes auricularis.

**Symptoms.**—In ordinary zona there is often a slight prodromal period in which the patient feels ill, has moderate fever, and pain in the side, sometimes of such severity as to suggest pleurisy. On the third or fourth day the rash appears. The characteristic group of vesicles has a segmental distribution limited to one side of the body. One or more of the adjoining skin fields is usually affected. With involvement of the cervical, lumbar, or sacral ganglion the zonal or girdle form of the vesicular crop is naturally lost owing to the distortion of the skin fields from the growth of the limbs. The typical zonal form is only seen in involvement of the thoracic ganglia. Groups of vesicles are regularly arranged on the hyperæmic skin, at first filled with a clear or sometimes bloody serum, which later becomes purulent. The crop varies greatly, and the individual vesicles may be superficial, in which case they leave no scar, or they may be deep and in healing leave superficial scars. By far the most serious form is that seen in the upper division of the fifth. The fever may be high and the eruption very profuse with great swelling and much pain. Permanent disfigurement may follow the scarring.

It seems not improbable, as Chauffard suggests, that there may be extension of the disease from the posterior ganglia to the neighboring meninges as there may be pains down the spine, the girdle sensation, exaggerated knee-jerks, the Kernig sign, and lymphocytosis in the cerebro-spinal fluid.

**Complications.**—The most serious of these is that occasionally seen in ophthalmic zoster, when there is intense inflammation of the conjunctiva and cornea with consecutive panophthalmitis and destruction of the eye.

In a few cases the eruption becomes gangrenous. Swelling of the lymph glands has been noted, sometimes before the eruption. A bilateral distribution has occurred. A generalized herpes zoster is occasionally seen with a widespread vesicular rash on the face, neck, trunk, and thighs. A facial paralysis may develop during or after ophthalmic or cervical herpes. Swelling of the parotid gland on the same side may occur. In rare cases paralysis of the extremities has occurred. A most distressing feature is *post-zonal neuralgia*. After recovery from the herpes, hot burning sensations remain in the cutaneous distribution. In other instances, particularly in the aged, the pain persists and for years may be a terrible affliction. The victim may commit suicide.

**Treatment.**—Care should be taken to protect the vesicles; a one per cent. cocaine ointment with lanolin applied on lint gives relief to the pain. Carbolyzed vaseline, stearate of zinc, menthol ointment, or dusting powders may be used. The salicylates or analgesic drugs aid in relieving pain. Benefit has been reported from the use of 1 c. c. of pituitary extract. In very severe involvement of the ophthalmic division of the fifth nerve the greatest care should be taken to keep the conjunctiva clean. For the severe post-zonal neuralgia, injections into the spinal cord have been tried, and in cases of great severity the posterior nerve roots may be cut.

## K. GENERAL AND FUNCTIONAL DISEASES

### I. PARALYSIS AGITANS

(*Parkinson's Disease; Shaking Palsy*)

**Definition.**—A chronic affection of the nervous system, characterized by disturbance of certain automatic and associated movements, tremors, and rigidity. The globus pallidus mechanism is affected.

**Etiology.**—By no means uncommon, the disease affects men more than women. It rarely occurs under forty, but instances have been reported in which the disease began about the twentieth year. Direct heredity is rare, but the patients often belong to families in which there are other nervous affections. In some cases it may be caused by senile degeneration and arterio-sclerotic changes. Among exciting causes may be mentioned business worries and anxieties; in some instances the disease has followed directly upon severe mental shock or trauma. Cases are described after the specific fevers.

**Morbid Anatomy.**—There are changes in the efferent motor system of the globus pallidus mechanism. In the juvenile type there are atrophy and decrease in number of the large motor cells of the globus pallidus system. These are regarded as a primary atrophy (abiotrophy). In the globus pallidus system the large cells are motor and the small ganglia cells are inhibitory and co-ordinating. If this destructive lesion involves both types of cells in the caudate nucleus and putamen, the Vogt syndrome results, that is, double athetosis with spastic contractures and pseudo-bulbar palsy. If the caudate nucleus and lenticular nucleus are the seat of this destructive lesion there results progressive lenticular degeneration—Wilson's disease—that is the paralysis agitans syndrome with rigidity, tremor, clonic and tonic spasms and perhaps choreic and athetoid movements (Gowers' tetanoid chorea).

**Symptoms.**—The disease begins gradually, usually in one or other hand, and the tremor may be either constant or intermittent. With this may be associated weakness or stiffness. At first these symptoms may be present only after exertion. Although the onset is slow and gradual in nearly all cases, there are instances in which it sets in abruptly after fright or trauma. When well established the disease is very characteristic. The following are the prominent features.

**THE FACE—PARKINSON'S MASK.**—Even before the tremor begins the expressionless face, slow movement of the lips, the elevated eyebrows, and general facial immobility suggest the disease. When well developed it is the most characteristic—and pathetic—feature.

**TREMOR.**—This may be in the four extremities or confined to hands or feet; the head is not so commonly affected. The tremor is usually marked in the hands, and the thumb and forefinger display the motion made in the act of rolling a pill. At the wrist there are movements of pronation and supination and though less marked, of flexion and extension. The upper-arm muscles are rarely involved. In the legs the movement is most evident at the ankle-joint, and less in the toes than in the fingers. Shaking of the head is

less frequent, but does occur, and is usually vertical, not rotatory. The rate of oscillation is about five per second. Any emotion exaggerates the movement. The attempt at a voluntary movement may check the tremor (the patient may be able to thread a needle), but it returns with increased intensity. The tremors cease, as a rule, during sleep, but persist when the muscles are not in use. The writing of the patient is tremulous and zigzag. For months or years the chief tremor may be in one arm or one leg.

**WEAKNESS.**—Loss of power is present in all cases, and may occur before the tremor, but is not very striking, as tested by the dynamometer, until the late stages. The weakness is greatest where the tremor is most developed. The movements are remarkably slow. There is rarely complete loss of power.

**RIGIDITY** may early be expressed in a slowness and stiffness in the voluntary movements, which are performed with some effort and difficulty, and all the actions of the patient are deliberate. This rigidity is in all the muscles, and leads ultimately to the characteristic attitude.

**ATTITUDE AND GAIT.**—The head is bent forward, the back bowed, and the arms held away from the body, somewhat flexed at the elbow-joints. The fingers are flexed and in the position assumed when the hand is at rest; in the late stages they can not be extended. Occasionally there is overextension of the terminal phalanges. The hand is usually turned toward the ulnar side and the attitude somewhat resembles that of advanced cases of arthritis deformans. In the late stages there are contractures at the elbows, knees, and ankles. The movements of the patient are characterized by great deliberation. He rises from the chair slowly in the stooping attitude, with the head projecting forward. In attempting to walk the steps are short and hurried, and, as Trousseau remarks, he appears to be running after his centre of gravity. This is termed festination or propulsion, in contradistinction to a peculiar gait observed when the patient is pulled backward, when he makes a number of steps and would fall over if not prevented—retropulsion.

The *voice*, as pointed out by Buzzard, is at first shrill and piping, and there is often a hesitancy in beginning a sentence; then the words are uttered with rapidity, as if the patient was in a hurry.

The **REFLEXES** are normal in most cases, but in a few they are exaggerated.

Of **SENSORY** disturbances Charcot noted alterations in the temperature sense. Some patients complain of subjective sensations of heat, general or local—which may be present on one side only and associated with an increase of the surface temperatures. In other instances, patients complain of cold. Localized sweating may be present. The skin, especially of the forehead, may be thickened. The mental condition rarely shows any change.

The disease is incurable. Periods of improvement may occur, but the tendency is for the affection to proceed progressively downward. It is a slow, degenerative process which lasts for years.

**VARIATIONS IN THE SYMPTOMS.**—The tremor may be absent, but the rigidity, weakness, and attitude are sufficient to make the diagnosis. The disease may be hemiplegic in character, involving only one side or even one limb. Usually these are but stages of the disease.

**Diagnosis.**—In well-developed cases the disease is recognized at a glance. The attitude, gait, stiffness, and mask-like expression are points of as much importance as the oscillations, and usually serve to separate the cases from

senile and other forms of tremor. Disseminated sclerosis develops earlier, and is characterized by the nystagmus and the scanning speech, and does not present the *attitude* so constant in paralysis agitans. Yet Schultze and Sachs have reported cases in which the signs of multiple sclerosis have been associated with those of paralysis. The hemiplegic form might be confounded with post-hemiplegic tremor, but the history, mode of onset, and greatly increased reflexes distinguish the two. The Parkinsonian face and rigidity are of great importance in the diagnosis of the obscure forms. The history should distinguish the Parkinsonian sequel of epidemic encephalitis.

**Treatment.**—There is no method which can be recommended as satisfactory in any respect. Slowly performed muscular movements, with strong mental concentration, are sometimes useful in controlling the tremor. Arsenic, opium and parathyroid gland extract (gr.  $\frac{1}{10}$ , 0.006 gm.) may be tried and sometimes give relief, but are not curative. Hyoscine seems helpful in some cases (gr.  $\frac{1}{200}$ , 0.0003 gm. and gradually increased).

#### OTHER FORMS OF TREMOR

**Simple Tremor.**—This is occasionally found in persons in whom it is impossible to assign any cause. It may be transient or persist for an indefinite time. It is often extremely slight, and is aggravated by all causes which lower the vitality.

**Hereditary Tremor.**—C. L. Dana reported remarkable cases of hereditary tremor. It occurred in all the members of one family, and beginning in infancy continued without producing any serious changes.

**Senile Tremor.**—With advancing age tremulousness during muscular movements is extremely common, but is rarely seen under seventy. It is always a fine tremor, which begins in the hands and often extends to the muscles of the neck, causing slight movement of the head.

**Toxic tremor** is seen chiefly as an effect of tobacco, alcohol, lead, or mercury; more rarely in arsenical or opium poisoning. In elderly men who smoke much it may be entirely due to tobacco. One of the commonest forms is the alcoholic tremor, which occurs only on movement and has considerable range. Lead tremor is considered under lead poisoning, of which it constitutes a very important symptom.

**Hysterical tremor**, which usually occurs under circumstances which make the diagnosis easy, will be considered in the section on hysteria.

## II. ACUTE CHOREA

(*Sydenham's Chorea; St. Vitus's Dance*)

**Definition.**—A disease, probably an acute infection, chiefly affecting children, characterized by irregular, involuntary contraction of the muscles, a variable amount of psychical disturbance, and a remarkable liability to acute endocarditis.

**Etiology.**—**SEX.**—Of 554 cases analyzed at the Philadelphia Infirmary for Nervous Diseases, 71 per cent. were in females and 29 per cent. in males

(Osler). Of 808 Johns Hopkins Hospital cases, 71.2 per cent. were females (Thayer and Thomas).

**AGE.**—The disease is most common between the ages of five and fifteen. Of 522 cases, 380 occurred in this period; 84.5 per cent. in Thayer and Thomas' series. It is rare among the negroes and native races of America. Only 25 of the Johns Hopkins Hospital cases were in negroes.

**RHEUMATIC FEVER.**—Of the 554 cases, in 15.5 per cent. there was a history of "rheumatism" in the family. In 88 cases, 15.8 per cent., there was a history of articular swelling, acute or subacute. In 33 cases there were pains, sometimes described as "rheumatic," in various parts, but not associated with joint trouble. Adding these to those with manifest articular trouble, the percentage is raised to nearly 21. It is rather remarkable that in the Baltimore series the percentage with a history of rheumatism was the same—21.6.

In one group the arthritis antedates by months or years the onset of the chorea, and does not recur before or during the attack. In the other the chorea sets in with or follows immediately upon the acute arthritis. It is difficult to differentiate the cases of irregular pains without definite arthritis. It is probable that many of them are rheumatic, but it is a mistake to regard as such all cases in children in which there are complaints of vague pains in the bones or muscles—so-called growing pains. It should never be forgotten that there may be no acute arthritis with rheumatic fever in a child.

**HEART DISEASE.**—Endocarditis is believed by some writers to be the cause of the disease. On this view chorea is the result of an embolic process occurring in the course of a rheumatic endocarditis.

**INFECTIOUS DISEASES.**—Scarlet fever with arthritic manifestations may be a direct antecedent. With the exception of rheumatic fever, there is no intimate relationship between chorea and the acute diseases incident to childhood. It may be noted in contrast to this that the so-called canine chorea is a common sequel of distemper. Chorea may follow gonorrhœa, puerperal fever, and other forms of sepsis. The tonsils are frequently diseased.

**SYPHILIS.**—There is a small group, with features much like those of chorea, in which congenital syphilis is apparently the cause. Specific treatment results in rapid improvement.

**ANÆMIA** is less often an antecedent than a sequence, and though cases occur in children who are anæmic and in poor health, this is by no means the rule. Chorea may come on during chlorosis.

**PREGNANCY.**—A choreic patient may become pregnant; more frequently the disease occurs during pregnancy; sometimes after delivery. Buist, of Dundee, tabulated 226 cases; in 6 the chorea preceded and in 105 it occurred during the pregnancy; in 31 in recurrent pregnancies; 45 cases terminated fatally, and in 16 cases the attack developed post partum. The alleged frequency in illegitimate primiparæ is not borne out by his figures. Beginning in the first three months were 108 cases, in the second three months 70 cases, in the last three months 25 cases. The disease is often severe, and maniacal symptoms may occur.

A tendency to the disease is found in certain families. In 80 cases there was a history of attacks of chorea in other members. In one instance both mother and grandmother had been affected. High-strung, excitable, nervous children are especially liable. *Fright* is considered a frequent cause, but in

a large majority of the cases no close connection exists between the fright and the onset of the disease. Occasionally the attack sets in at once. Mental worry, trouble, a sudden grief, or a scolding may apparently be the exciting cause. The strain of *education*, particularly in girls during the third hemidecade, appears to be an important factor. Bright, intelligent, active-minded girls from ten to fourteen, ambitious to do well at school, often stimulated in their efforts by teachers and parents, form a large contingent of the cases—the so-called *school-made chorea*. *Imitation*, which is mentioned as an exciting cause, is extremely rare, and did not appear to have influenced the onset in a single case in the Infirmary records.

The disease may rapidly follow an injury or a slight surgical operation. Reflex irritation was believed to play an important rôle, particularly the presence of worms or genital irritation, but this is very doubtful. Ocular defects do not occur in greater proportion in choreic than in other children; and a majority of the cases in which operation has been followed by relief have been instances of *tic*, local or general.

**Pathology.**—Two anatomical changes are found: (1) *Endocarditis*, usually simple (and of the mitral valve), which was present in 62 of 73 fatal cases recorded.<sup>1</sup> In a few instances the lesion was ulcerative. (2) Foci of softening in the *basal ganglia*, in the situation and with the appearance of an *acute encephalitis*. Minute hæmorrhages have been found elsewhere in the brain. Connected with the endocarditis there are on record seven cases of embolism of the central artery of the retina (H. M. Thomas) and cerebral embolism has been found.

The *pathology* is still obscure. That it is an acute infection is suggested by (1) the association with rheumatic fever; (2) the character of the acute febrile cases; (3) the frequency of involvement of the tonsils; (4) the seasonal relations; (5) the presence of endocarditis; (6) the finding of micro-organisms—though the *Diplococcus rheumaticus* is not generally accepted as the cause; and (7) the occurrence of a chorea type in epidemic encephalitis in which the lesions are very similar to and in the same situation, basal ganglia, as in simple chorea.

**Symptoms.**—Three groups of cases may be recognized—the mild, severe, and maniacal chorea.

*Mild Chorea.*—In this the affection of the muscles is slight, the speech is not seriously disturbed, and the general health not impaired. Premonitory symptoms are shown in restlessness and inability to sit still, a condition well characterized by the term “fidgets.” There are emotional disturbances, such as crying spells, or sometimes night terrors. There may be pains in the limbs and headache. Digestive disturbances and anæmia may be present. A change in the temperament is frequently noticed, and a docile, quiet child may become cross and irritable. After these symptoms have persisted for a week or more the characteristic involuntary movements begin, and are often first noticed at the table, when the child spills a tumbler of water or upsets a plate. There may be only awkwardness or slight incoördination of voluntary movements, or constant irregular clonic spasms. The jerky, irregular character of the movements differentiates them from almost every other disorder.

<sup>1</sup>Osler, *Chorea and Choreiform Affections*, Philadelphia, 1894.

of motion. In the mild cases only one hand, or the hand and face, are affected, and it may not spread to the other side.

In the *severe form* the movements become general and the patient may be unable to get about or to feed or undress herself, owing to the constant, irregular, clonic muscular contractions. The speech is affected, and for days the child may not be able to talk. Often with the onset of the severer symptoms there is loss of power on one side or in the limb most affected.

The third and most extreme form, maniacal chorea or *chorea insaniens*, is truly a terrible disease, and may arise out of the ordinary form. These cases are more common in adult women and may develop during pregnancy.

Chorea begins, as a rule, in the hands and arms, then involves the face, and subsequently the legs. The movements may be confined to one side—*hemi-chorea*. The attack begins oftenest on the right side, though occasionally it is general from the outset. One arm and the opposite leg may be involved. In a number of cases speech is affected; this may amount only to an embarrassment or hesitancy, but in other instances it becomes an incoherent jumble. In very severe cases the child will make no attempt to speak, perhaps for weeks. This is not marked by special choreic unrest of the muscles of speech; it is probably a motor weakness. Complete recovery follows. Paroxysms of panting and of hard expiration may occur, or odd sounds may be produced. As a rule the movements cease during sleep.

*Weakness*.—A prominent symptom is muscular weakness, usually no more than a condition of paresis. The loss of power is slight, but the weakness may be shown by an enfeebled grip or by a dragging of the leg or limping. In some cases there is flaccidity of the limbs (*limp chorea*). In his original account Sydenham refers to the “unsteady movements of one of the legs, which the patient drags.” There may be extreme paresis with but few movements—the paralytic chorea of Todd. Occasionally a local paralysis or weakness remains after the attack.

*CARDIAC*.—As so many of the subjects of chorea are nervous girls, it is not surprising that a rapidly acting heart is common. Irregularity is not so special a feature. The patients seldom complain of pain about the heart.

*Murmurs*.—With anæmia and debility, not uncommon associates of chorea in the third or fourth week, we find a corresponding cardiac condition. The impulse is diffuse, perhaps wavy in thin children. The carotids throb visibly, and in the recumbent posture there may be pulsation in the cervical veins. On auscultation a systolic murmur is heard at the base, perhaps, too, at the apex, soft and blowing in quality.

*Endocarditis*.—Acute valvulitis rarely gives evidence of its presence by symptoms. It must be sought, and it is usually associated with murmurs at one or other of the cardiac orifices.

For the guidance of the practitioner these statements may be made:

(a) In thin, nervous children a systolic murmur of soft quality is extremely common at the base, with accentuation of the second sound, particularly at the second left costal cartilage, and is probably of no moment.

(b) A systolic murmur of maximum intensity at the apex, and heard also along the left sternal margin, is not uncommon in anæmic, enfeebled states, and does not necessarily indicate either endocarditis or insufficiency.

(c) A murmur of maximum intensity at the apex, with rough quality, and

transmitted to the axilla or angle of the scapula, indicates an organic lesion of the mitral valve, and is usually associated with enlargement of the heart.

(d). When in doubt it is much safer to trust to the evidence of eye and hand than to that of the ear. If the apex beat is in the normal position, and the area of dullness not increased vertically or to the right of the sternum, there is probably no serious valvular disease.

The *endocarditis* of chorea is almost invariably of the simple form, and in itself not dangerous; but it leads to those sclerotic changes in the valve which produce incompetency. Of 140 patients examined more than two years after the attack, the heart was normal in only 51; in 17 there was functional disturbance, and 72 presented signs of organic heart disease. In an analysis of the Johns Hopkins Hospital cases, Thayer found evidence of involvement of the heart in 25 per cent. of the out-patients and in more than 50 per cent. of the ward patients. Cardiac involvement was more common in the cases with a history of rheumatic fever, and was much more frequent in the relapses. Pericarditis is an occasional complication.

SENSORY DISTURBANCES.—Pain in the affected limbs is not common. Occasionally there is soreness on pressure. There are cases, usually of hemichorea, in which pain in the limbs is marked. Weir Mitchell spoke of these as *painful choreas*. Tender points along the lines of emergence of the spinal nerves or along the course of the nerves of the limbs are rare.

PSYCHICAL DISTURBANCES are common. Irritability of temper, marked wilfulness, and emotional outbreaks may indicate a complete change in the character. There is deficiency in the powers of concentration, the memory is enfeebled, and the aptitude for study is lost. The psychical element is apt to be neglected and it is always a good plan to tell the parents that it is not the muscles alone which are affected, but that the irritability and change of disposition really form part of the disease. Rarely there is progressive impairment of the intellect with termination in actual dementia. Acute melancholia has been described. Hallucinations of sight and hearing may occur. Patients may behave in an odd manner and do all sorts of meaningless acts. The most serious manifestation of this character is the maniacal delirium, occasionally associated with the very severe cases—*chorea insaniens*. Usually the motor disturbance in these cases is aggravated, but it has been overlooked and patients have been sent to an asylum.

The deep *reflexes* often show much variation, especially the knee-jerk; the quadriceps contraction may be unusually prolonged. Trophic lesions rarely occur unless, as some writers have done, the joint troubles are regarded as arthropathies occurring in the course of a cerebro-spinal disease.

FEVER, usually slight, was present in all but one of 110 cases (Thayer). Endocarditis may occur with little if any rise in temperature; but, on the other hand, with an acute arthritis, severe endocarditis or pericarditis, and in the maniacal cases the fever may range from 102° to 104°.

CUTANEOUS AFFECTIONS.—The pigmentation, which is not uncommon, is due to the arsenic. Herpes zoster occasionally occurs. Erythema nodosum and a purpuric urticaria have been described. There may, indeed, be the more aggravated condition of rheumatic purpura, known as Schönlein's *peliosis rheumatica*.

Subcutaneous fibrous nodules may be present.

**Course.**—From eight to ten weeks is the average duration of an attack of moderate severity. Cases described as chronic chorea following an acute attack are usually instances of cerebral sclerosis or Friedreich's ataxia; but occasionally an attack which has come on in the ordinary way persists for months or years, and recovery ultimately takes place. A slight grade, particularly noticeable under excitement, may persist for months in nervous children. The tendency to *recur* has been noticed by all writers since Sydenham first made the observation. Of 410 cases analyzed for this purpose, 240 had one attack, 110 had two attacks, 35 three attacks, 10 four attacks, 12 five attacks, and 3 six attacks. The recurrence is apt to be in the spring.

Recovery is the rule. The statistics of outpatient departments are not favorable for determining the mortality. A reliable estimate is that of the Collective Investigation Committee of the British Medical Association, in which 9 deaths were reported among 439 cases, about 2 per cent. There were 141 deaths in the United States registration area in 1920.

The paralysis rarely persists. Mental dulness may be present for a time, but usually passes away; permanent impairment of the mind is exceptional.

**Diagnosis.**—In a majority of instances the diagnosis is made at a glance; but there are several affections which may be mistaken for it.

(a) *Multiple and Diffuse Cerebral Sclerosis.*—The cases are often mistaken for ordinary chorea, and have been described as *chorea spastica*. As a rule, the movements are readily distinguishable from those of true chorea, but the simulation is sometimes close; the onset in infancy, impaired intelligence, increased reflexes and in some instances rigidity with the chronic course separate them sharply from true chorea.

(b) *Friedreich's Ataxia.*—Cases of this well-characterized disease were formerly classed as chorea. The slow, irregular, incoördinate movements, the scoliosis, the scanning speech, the early talipes, the nystagmus, and the family character of the disease are points which render the diagnosis easy.

(c) In rare cases the paralytic form of chorea may be mistaken for *poliomyelitis* or, when both legs are affected, for paraplegia of spinal origin; but this can be the case only when the choreic movements are very slight.

(d) *Hysteria* may simulate chorea more closely, and unless there are other manifestations it may be impossible to make a diagnosis. Most commonly, however, the movements in the so-called hysterical chorea are rhythmic and differ entirely from those of ordinary chorea.

(e) The *mental symptoms* in maniacal chorea may mask the true nature of the disease especially if the choreic movements disappear.

(f) *Habit spasms* and *tics* should not be confused.

(g) *Epidemic encephalitis* with chorea-like movements is recognized by the other features of the disease.

**Treatment.**—Abnormally bright, active-minded children belonging to families with pronounced neurotic taint should be carefully watched and not allowed to overtax their mental powers. So frequently in children of this class does the attack of chorea date from the worry and stress incident to examinations that the competition for prizes should be emphatically forbidden.

The treatment of the attack consists largely in attention to hygienic measures, with which alone, in time, a majority of the cases recover. Parents should be told to scan gently the faults and waywardness of choreic children.

The psychical element, strongly developed in so many cases, is best treated by quiet and seclusion. The child should be confined to bed in the recumbent posture, and mental as well as bodily quiet enjoined. In private practice this is often impossible, but with well-to-do patients the disease is always serious enough to demand the assistance of a skilled nurse. Toys and dolls should not be allowed at first, for the child should be kept amused without excitement. The rest allays the hyper-excitability and reduces to a minimum the possibility of damage to the valve segments should endocarditis exist.

The child should be kept apart from other children and, if possible, from other members of the family, and should see only those persons directly concerned with the care and nursing. The child should be protected against chilling. If the movements are violent care should be taken to prevent the child from falling out of bed or injuring herself.

**Diet.**—Every effort should be made to give sufficient nourishment. Many patients do well on a diet principally of milk to which lactose may be added. Fruit juices, cooked fruits, ice cream and cocoa are allowed. Soft foods may be given if desired. The nasal tube should be used if there is difficulty in swallowing.

**Medicinal.**—The salicylates may be used for fever; acetyl salicylic acid, (gr. 5-10, 0.3-0.6 gm.) is often the best. The use of arsenic is very general but it is a question if very large doses are indicated. It may be given as Fowler's solution beginning with 5 minims (0.3 c. c.) three times a day and increasing to double this amount. It should be stopped at once if there are any signs of overdosage. *Sedatives* are usually necessary. Chloral hydrate is usually the best, the required dosage varying greatly. It may be begun in 10 grain (0.6 gm.) doses three or four times a day and increased if necessary. An equal amount of bromide may be added. Belladonna is useful in some cases. Hyosine hydrobromide (gr. 1/100, 0.0006 gm.) sometimes is helpful. Barbitol or pheno-barbitol in suitable dosage to the age is useful in securing sleep. In case of congenital syphilis active specific treatment should be given. Simple laxatives or enemata are used to keep the bowels regular. For anemia, iron and arsenic preparations are indicated.

Electricity is of doubtful value. The question of gymnastics is an important one. Early in the disease, when the movements are active, they are not advisable; but during convalescence carefully graduated exercises are beneficial. It is not well to send a choreic child to a school gymnasium, as the stimulus of other children and the excitement are very prejudicial.

In the severe cases with incessant movements, sleeplessness, dry tongue, and delirium, the important indication is to procure rest, for which purpose chloral may be freely given, and, if necessary, morphia. Chloroform inhalations may be necessary to control the intensity of the paroxysms, but the high rate of mortality in this form illustrates how often our endeavors are fruitless. The wet pack is sometimes soothing and should be tried. As these patients are apt to sink rapidly into a low typhoid state with myocardial weakness, a supporting treatment is required from the outset.

There are cases which drag on from month to month without getting better or worse, and resist all modes of treatment. In such cases a combination of suggestion and passive movements, followed by voluntary movements under control, and later simple exercises, may be useful. Change of air and scene

is sometimes followed by rapid improvement, and in these cases the treatment by rest and seclusion should always be given a full trial.

Diseased tonsils should be removed and nasal trouble corrected. Marked ocular defects should be properly corrected. After the child has recovered, the parents should be warned that return is by no means infrequent, and is particularly liable to follow overwork at school or debilitating influences. Sydenham advised purging to prevent the recurrence.

### III. HABIT SPASMS AND TICS

**Habit Spasm; Convulsive Tic.**—Two groups of cases may be recognized under the designation of habit spasm—one in which there are simply localized spasmodic movements, and the other in which, in addition to this, there are explosive utterances and psychical symptoms, a condition to which French writers have given the name *tic convulsif*.

(a) **HABIT SPASM.**—This is found chiefly in childhood, most frequently in girls from seven to fourteen years of age (Mitchell). There is usually a psychical basis; imitation is a factor in some cases. In its simplest form there is a sudden, quick contraction of certain of the facial muscles, such as rapid winking or drawing of the mouth to one side, or the neck muscles are involved and there are unilateral movements of the head. The head is given a sudden, quick shake, and at the same time the eyes wink. A not infrequent form is the shrugging of one shoulder. The grimace or movement is repeated at irregular intervals, and is much aggravated by emotion. A short inspiratory sniff is not an uncommon symptom. Night terrors and enuresis may be associated. The cases are most frequent in children who are "out of sorts," who have been growing rapidly, or who have inherited a tendency to neurotic disorders. Allied to or associated with this are some of the curious tricks of children. A boy was in the habit every few moments of putting the middle finger into the mouth, biting it, and at the same time pressing his nose with the forefinger. Hartley Coleridge is said to have had a somewhat similar trick, only he bit his arm. In all these cases the habits of the child should be examined carefully, the nose and vault of the pharynx thoroughly inspected, and the eyes tested. As a rule the condition is transient, and after a few months gradually disappears. Occasionally a local spasm persists—twitching of the eyelids or the facial grimace.

*Spasmus nutans*, head nodding, is a co-ordinated tic in young infants usually of a harmless nature; it may be associated with nystagmus.

In *treatment*, comments on the condition and the mimicry of other children should be avoided. The child should lead a quiet open-air life with sufficient exercise and amusement. A tactful effort should be made to gain the child's confidence and learn his mental processes. The origin of the habit may be due to some curious idea or fear. In severe acute cases a period of rest and seclusion may be advisable. In older children and adults the effort should be made to learn to relax the muscles concerned. If there is much nervous excitement bromides are useful.

(b) **IMPULSIVE TIC (GILLES DE LA TOURETTE'S DISEASE).**—This remarkable affection, often mistaken for chorea, more frequently for habit spasm, is

really a psychosis allied to hysteria; psychical tic, though in certain of its aspects it has the features of monomania. The disease begins, as a rule, in young children, occurring as early as the sixth year, though it may occur after puberty. There is usually a neurotic family history. The special features are:

(1) Involuntary muscular movements, usually affecting the facial or brachial muscles, but in aggravated cases all the muscles of the body may be involved and the movements may be extremely irregular and violent.

(2) Explosive utterances, which may resemble a bark or an inarticulate cry. A word heard may be mimicked at once and repeated over and over again, usually with the involuntary movements. To this the term *echolalia* has been applied. A much more distressing disturbance in these cases is *coprolalia*, or the use of bad language. A small child may shock its mother and friends by constantly swearing when making the involuntary movements or by uttering all sorts of obscene words. Occasionally actions are mimicked—*echokinesis*.

(3) Associated with some of these cases are curious mental disturbances; the patient becomes the subject of a form of obsession or a fixed idea. This may take the form of the impulse to touch objects, or it is a fixed idea about words—onomatomania—or the patient may feel compelled to count a number of times before doing certain actions—arithmomania. The disease is readily distinguished from ordinary chorea. The movements have a larger range and are explosive in character. Tourette regards the coprolalia as the most distinctive feature. The prognosis is doubtful, but recovery may follow. In treatment every effort should be made to gain the patient's confidence and study his psychical processes, early in the course if possible. Psychical treatment is the most important. His life should be ordered in the most healthy fashion, physically and mentally.

**Saltatory Spasm** (*Latah*; *Myriachit*; *Jumpers*).—Bamberger has described a disease in which when the patient attempted to stand there were strong contractions in the leg muscles, which caused a jumping or springing motion. This occurs only when the patient attempts to stand. The affection has occurred in both men and women, more frequently in the former, and the subjects have usually shown marked neurotic tendencies. In many cases the condition has been transitory; in others it has persisted for years. Remarkable affections similar to this in certain points occur as a sort of epidemic neurosis. One of the most striking of these occurs among the "jumping Frenchmen" of Maine and Canada. As described by Beard and Thornton, the subjects are liable on any sudden emotion to jump violently and utter a loud cry or sound, and will obey any command or imitate any action without regard to its nature. The condition of echolalia is present in a marked degree. The "jumping" prevails in certain families.

A very similar disease prevails in parts of Russia and in Java and Borneo, where it is known by the names of myriachit and latah, the chief feature of which is mimicry by the patient of everything he sees or hears.

**Rhythmic Chorea**.—This is readily recognized by the rhythmical character of the movements. It may affect the muscles of the abdomen, producing the salaam convulsion, or involve the sterno-mastoid, producing a rhythmical movement of the head, or the psoas, or any group of muscles. In its orderly

rhythm it resembles the canine chorea.\* It has to be distinguished from the myoclonic form of epidemic encephalitis.

#### IV. INFANTILE CONVULSIONS

Convulsive seizures similar to those of epilepsy are not infrequent in children. The fit may be identical with epilepsy, from which the condition differs in that when the cause is removed there is no tendency for the fits to recur. Occasionally, however, the convulsions continue and pass into true epilepsy.

**Etiology.**—A convulsion may be due to many causes, all of which lead to an unstable condition of the nerve centres, permitting sudden, excessive, and temporary nervous discharges. The following are the most important:

(1) *Debility*, resulting usually from gastro-intestinal disturbance. Convulsions frequently supervene toward the close of an attack of entero-colitis and recur, sometimes proving fatal. The death-rate in children from eclampsia rises steadily with that of gastro-intestinal disorders (M. J. Lewis).

(2) *Irritation*.—Dentition alone is rarely a cause, but is often one of several factors in a feeble, unhealthy infant. The greatest mortality from convulsions is during the first six months, before the teeth have really cut through the gums. Other irritative causes are the overloading of the stomach with indigestible food. It has been suggested that some of these cases are toxic. Worms, to which convulsions are frequently attributed, probably have little influence. Among other sources possible are phimosia and otitis.

(3) *Rickets*.—Rickets and convulsions are often associated (Jenner.) The spasms may be laryngeal, the so-called child-crowing, which, though convulsive in nature, can scarcely be reckoned under eclampsia. The influence of this condition is more apparent in Europe than in the United States, although rickets is a common disease, particularly among the colored people. Spasms, local or general, in rickets are probably associated with the condition of debility and malnutrition, and with craniotabes.

(4) *Fever*.—In young children the onset of the infectious diseases is frequently with convulsions, which may take the place of a chill in the adult. It is not known upon what they depend. Scarlet fever, measles, and pneumonia are most often preceded by convulsions.

(5) *Congestion of the Brain*.—That extreme engorgement of the blood-vessels may produce convulsions is shown by their occasional occurrence in severe whooping-cough, but their rarity in this disease really indicates how small a part mechanical congestion plays in the production of fits.

(6) *Severe convulsions* usher in or accompany many of the serious diseases of the nervous system in children. The acute encephalitis of children, which is followed by hemiplegia, usually has severe convulsions at the onset. They less frequently precede a spinal paralysis. They occur with meningitis, tuberculous or simple, and with tumors and other lesions of the brain.

And, lastly, convulsions may occur immediately after birth and persist for weeks or months. In such instances there has probably been meningeal hæmorrhage or serious injury to the cortex.

The relation of convulsions in children to true *epilepsy* is important. In Gowers' figures of 1,450 cases of epilepsy, the attacks began in 180 during the

first three years of life. Of 460 cases of epilepsy in children, in 187 the fits began within the first three years and the greatest number, 74, was in the first year (Osler). In nearly all these instances there was no interruption in the convulsions. J. L. Morse regards as the dangerous forms those in which the convulsions occur over a considerable period or in which there are repeated attacks suggesting *petit mal*.

**Symptoms.**—The attack may come on suddenly without any warning; more commonly it is preceded by restlessness, twitchings and perhaps grinding of the teeth. The convulsion is rarely so complete in its stages as true epilepsy. The spasm begins usually in the hands, most commonly in the right hand. The eyes are fixed and staring or are rolled up. The body becomes stiff and breathing is suspended for a moment or two by tonic spasm of the respiratory muscles, in consequence of which the face becomes congested. *Clonic* convulsions follow, the eyes are rolled about, the hands and arms twitch, or are fixed and extended in rhythmical movements, the face is contorted, and the head is retracted. The attack gradually subsides and the child sleeps or passes into a state of stupor. Following indigestion the attack may be single, but in rickets and intestinal disorders it is apt to be repeated. Sometimes the attacks follow each other with great rapidity, so that the child never rouses but dies in a deep coma. If the convulsion has been limited chiefly to one side there may be slight paresis after recovery, or if the convulsions usher in infantile hemiplegia, when the child arouses, one side is completely paralyzed. During the fit the temperature is often raised. Death rarely occurs from the convulsion itself, except in debilitated children or when the attacks recur with great frequency. In the so-called hydrocephaloid state in connection with protracted diarrhoea convulsions may close the scene.

**Diagnosis.**—If the subject is in full health, the attack is probably due to an overloaded stomach, to some peripheral irritation, or occasionally to trauma. Setting in with high fever and vomiting, it may indicate the onset of an exanthem, meningitis, encephalitis, or whatever the condition is which causes infantile hemiplegia. When the attack is associated with debility or rickets the diagnosis is easily made. The carpopedal spasms and pseudo-paralytic rigidity which are often associated with rickets, laryngismus stridulus, and the hydrocephaloid state are usually confined to the hands and arms and are intermittent and usually tonic. The convulsions associated with tumor or those which follow infantile hemiplegia are usually at first Jacksonian in character. After the second year convulsive seizures which come on irregularly without apparent cause and recur while the child is apparently in good health, are likely to prove true epilepsy.

**Prognosis.**—Convulsions play an important part in infantile mortality. In chronic diarrhoea convulsions are usually of ill omen. Those ushering in fevers are rarely serious, and the same may be said of the fits associated with indigestion and peripheral irritation.

**Treatment.**—Every source of irritation should be removed. If associated with indigestible food, a prompt emetic should be given, followed by an enema. The teeth should be examined, and if the gum is swollen, hot, and tense, it may be lanced; but never if it looks normal. When seen at first, if the paroxysm is severe, no time should be lost by giving a hot bath, but chloroform should be given at once, and repeated if necessary. A child is so readily put

under chloroform and with such a small quantity that this procedure is quite harmless and saves valuable time. The practice is almost universal of putting the child into a warm bath, and if there is a fever the head may be doused with cold water. The temperature of the bath should not be above 95° or 96°. The very hot bath is not suitable, particularly if the fits are due to indigestion. After the attack an ice-cap may be placed upon the head. If there is much irritability, particularly in rickets and severe diarrhœa, small doses of opium will be found efficacious. When the convulsions recur after the child comes from under the influence of chloroform it is best to place it rapidly under the influence of opium, which may be given as morphia hypodermically, in doses of gr. 1/25 to 1/30 (0.0026 to 0.0022 gm.) for a child of one year. Other remedies are chloral by enema, in 5 grain (0.3 gm.) doses, and nitrite of amyl. After the attack has passed the bromides are useful, of which 5 to 8 grains (0.3 to 0.5 gm.) may be given in a day to a child a year old. Recurring convulsions, particularly if they come on without special cause, should receive careful treatment with bromides. When associated with rickets the treatment should be directed to this disease.

## V. EPILEPSY

**Definition.**—An affection of the nervous system characterized by attacks of unconsciousness, with or without convulsions. The transient loss of consciousness without convulsive seizures is known as minor epilepsy (*petit mal*); the loss of consciousness with general convulsive seizures is known as major epilepsy (*grand mal*). Localized convulsions, occurring usually without loss of consciousness, are known as Jacksonian or cortical epilepsy.

**Etiology.**—Idiopathic or essential epilepsy, the form with an unknown or indefinite etiology, appears to depend upon a congenital tendency in the individual. Coarse anatomical changes in the brain are not present, but with the development of technique alterations have been determined in an increasing proportion of cases, particularly a gliosis of the superficial layers of the cortex described by Alzheimer. Apart from this is the large group of *symptomatic* convulsive seizures due to *toxæmias*, trauma, growths, chronic infections and arterio-sclerosis, which should not be termed epilepsy.

**AGE.**—In a large proportion of cases the disease begins before puberty. Of 1,450 cases observed by Gowers, in 422 the disease began before the tenth year, and three-fourths of the cases began before the twentieth year. Of 427 cases of epilepsy in children, the age of onset was as follows: First year, 74; second year, 62; third year, 51; fourth year, 24; fifth year, 17; sixth year, 18; seventh year, 19; eighth year, 23; ninth year, 17; tenth year, 27; eleventh year, 17; twelfth year, 18; thirteenth year, 15; fourteenth year, 21; fifteenth year, 34. Arranged in hemidecades the figures are as follows: From the first to the fifth year, 229; from the fifth to the tenth year, 104; from the tenth to the fifteenth year, 95 (Osler). These figures illustrate in a striking manner the early onset in a large proportion of cases. It is well always to be suspicious of "epilepsy" beginning in adult life, for in a majority of such cases the convulsions are due to a local lesion.

**Sex.**—No special influence is evident, certainly not in children. Of 435 cases, 232 were males and 203 were females. After puberty unquestionably, if a large number of cases are taken, the males are in excess.

**HEREDITY.**—Gowers remarks "there are few diseases in the production of which inheritance has a more marked influence." Of 2,523 epileptics, 16 per cent. were due to heredity (Spratling). The study of the American Eugenics Bureau (Bulletin No. IV), analyzing data of 206 epileptics, shows how potent are inherited factors. Pierce Clark considers that there are more or less definite essential defects in epileptics which account in part for the predisposition. These are "egocentricity, supersensitiveness, an emotional poverty and an inherent lack of adaptability to normal social life." Stress and annoyance, and an intensive regression to day-dreaming, lethargy and somnolence are precipitating factors. "The attack occurs at the final break of a too severe tension."

*Chronic alcoholism* in the parents is regarded by many as a potent predisposing factor. Echeverria analyzed 572 cases and divided them into three classes, of which 257 cases could be traced directly to alcohol as a cause; 126 cases in which there were associated conditions, such as syphilis and traumatism; 189 cases in which the alcoholism was probably the result of the epilepsy. Figures equally strong are given by Martin, who in 150 insane epileptics found 83 with a marked history of parental intemperance. Spratling found 15 per cent. with marked alcoholic history in the parents. Severe convulsive seizures may occur in steady drinkers.

*Syphilis.*—This in the parents is probably less a predisposing than an actual cause of epilepsy, which is the direct outcome of local cerebral manifestations. There is no reason for recognizing a special form of syphilitic epilepsy. On the other hand, convulsive seizures due to acquired syphilitic disease of the brain are very common.

Of exciting causes fright is probably not important. Trauma is present in a certain number of instances. An important group depends upon a local disease of the brain existing from childhood, as seen in the post-hemiplegic epilepsy. Occasionally cases follow the infectious fevers. Masturbation is stated to be a cause but its influence is overrated. A group of convulsive seizures like epilepsy is due to some toxic agent, as in lead poisoning and uræmia.

**REFLEX CAUSES.**—Eye strain, dentition, worms, the irritation of a cicatrix, some local affection, such as adherent prepuce, or a foreign body in the ear or nose, are given as causes. In rare cases the fits cease after the removal of the irritating factor but usually the attacks persist. Genuine cases of reflex epilepsy are rare. A remarkable instance occurred in a man with a testis in the inguinal canal, pressure upon which caused a typical fit. Removal of the organ was followed by cure.

Cardio-vascular "epilepsy" is usually a manifestation of advanced arteriosclerosis, and is associated with slow pulse. (See Stokes-Adams Disease.) The passage of a gall-stone or the removal of pleuritic fluid may induce a fit. Digestive troubles are extremely common in epilepsy and the eating of indigestible articles seems often to precipitate an attack. Protein hypersensitivity to certain foods is found in some patients. Convulsive seizures may occur in old people without obvious cause.

**Symptoms.**—(a) **MAJOR EPILEPSY.**—Preceding the fits there is usually a localized sensation, known as an *aura*, in some part of the body. This may be somatic, in which the feeling comes from some particular region in the periphery, as from the finger or hand, or is a sensation felt in the stomach or about the heart. The peripheral sensations are of great value, particularly those in which the aura always occurs in a definite region, as in one finger or toe. It is the equivalent of the signal symptom in a fit from a brain tumor. The varieties of these sensations are numerous. The epigastric sensations are most common. In these the patient complains of an uneasy sensation in the epigastrium or distress in the intestines, or the sensation may be not unlike that of heartburn and associated with palpitation. These groups are sometimes known as pneumogastric auræ or warnings.

Of *psychical* auræ one of the most common, as described by Hughlings Jackson, is a vague, dreamy state, a sensation of strangeness or sometimes of terror. The auræ may be associated with special senses; of these the most common are the visual, consisting of flashes of light or sensations of color; less commonly, distinct objects are seen. The auditory auræ consist of noises in the ear, odd sounds, musical tones, or occasionally voices. Olfactory and gustatory auræ, unpleasant tastes and odors, are rare.

Occasionally the fit may be preceded not by an aura, but by certain movements; the patient may turn round rapidly or run with great speed for a few minutes, the so-called *epilepsia procursiva*. In an Elwyn case the lad stood on his toes and twirled with extraordinary rapidity, so that his features were scarcely recognizable. It is stated that the pulse sometimes stops just before the fit. The studies of Gibson and Good show that no alteration in the pulse occurred up to the point of clonic convulsions, and there was no lowering of the blood pressure suggesting anæmia of the brain. At the onset of the attack the patient may give a loud scream or yell, the so-called *epileptic cry*. The patient drops as if shot, making no effort to guard the fall. In consequence, epileptics frequently injure themselves, cutting the face or head or burning themselves. In the attack, as described by Hippocrates, "the patient loses his speech and chokes, and foam issues from the mouth, the teeth are fixed, the hands are contracted, the eyes distorted, he becomes insensible, and in some cases the bowels are affected. And these symptoms occur sometimes on the left side, sometimes on the right, and sometimes on both." The fit may be described in three stages:

(1) *Tonic Spasm.*—The head is drawn back or to one side and the jaws are fixed. The hands are clinched and the legs extended. This tonic contraction affects the muscles of the chest, so that respiration is impeded and the initial pallor of the face changes to a dusky or livid hue. The muscles of the two sides are unequally affected, so that the head and neck are rotated or the spine is twisted. The arms are usually flexed at the elbows, the hand at the wrist, and the fingers are tightly clinched in the palm. This stage lasts only a few seconds, and then the clonic stage begins.

(2) *Clonic Stage.*—The muscular contractions become intermittent; at first tremulous or vibratory, they gradually become more rapid and the limbs are jerked and tossed about violently. The muscles of the face are in constant clonic spasm, the eyes roll, the eyelids are opened and closed convulsively. The movements of the muscles of the jaw are very forcible and strong, and

at this time the tongue is apt to be caught between the teeth and lacerated. The cyanosis, marked at the end of the tonic stage, gradually lessens. A frothy saliva, which may be blood stained, escapes from the mouth. The faeces and urine may be discharged involuntarily. The duration of this stage is variable. It rarely lasts more than one or two minutes. The contractions become less violent and the patient passes into the condition of coma.

(3) *Coma*.—The breathing is noisy or even stertorous, the face congested, but no longer intensely cyanotic. The limbs are relaxed and the unconsciousness is profound. After a variable time the patient can be aroused, but if left alone he sleeps for some hours and then awakes, complaining only of slight headache or mental confusion. If the attack has been severe, petechial hæmorrhages may occur over the neck and chest. In a young man in a severe convulsion both subconjunctival spaces were entirely filled with blood, and free blood oozed from them (Walter James). Hæmoptysis is a rare sequel.

After the attack the reflexes are often absent for a short time and later increased; ankle clonus can usually be obtained. The state of the urine is variable, particularly as regards the solids. The quantity is usually increased after the attack, and albumin or sugar may be present.

(4) *Status epilepticus*.—In this attacks occur in rapid succession, and the patient does not recover consciousness. The pulse, respiration, and temperature rise in the attack. It is a serious condition, and often proves fatal.

(5) *Post-epileptic symptoms* are of great importance. The patient may be in a trance-like condition, in which he performs actions of which subsequently he has no recollection. More serious are the attacks of mania, in which the patient is often dangerous and sometimes homicidal. It is held by some that an outbreak of mania may be substituted for the fit. And, lastly, the mental condition of an epileptic patient is often seriously impaired.

(6) *Paralysis*, which rarely follows the epileptic fit, is usually hemiplegic and transient. Slight disturbances of speech may occur; in some instances, forms of sensory aphasia. Scripture draws attention to an inflexibility of speech of the epileptic which sounds "expressionless or wooden" and can be recognized by a trained ear. The absence of flexibility can be demonstrated by graphic records.

The attacks may occur at night, and a person may be epileptic for years without knowing it. As Trousseau remarks, when a person tells us that in the night he has incontinence of urine, awakes in the morning with headache and mental confusion, complains of difficulty in speech because he has bitten his tongue, and there are purpuric spots on the face and neck, the probability is very strong that he is subject to nocturnal epilepsy.

(b) *MINOR EPILEPSY*.—Epilepsy without convulsions consists of transient unconsciousness, which may come on at any time, with or without a feeling of faintness and vertigo. Suddenly, at the dinner table, the subject stops talking and eating, the eyes become fixed, and the face slightly pale. Anything which may have been in the hand is usually dropped. In a moment or two consciousness is regained and the patient resumes conversation as if nothing had happened. In other instances there is slight incoherency or the patient performs some almost automatic action. He may begin to un-

dress and on returning to consciousness find that he has partially disrobed. He may rub his face, or spit about in a careless way. In other attacks the patient falls without convulsive seizures. A definite aura is rare. Though transient unconsciousness and giddiness are the most constant manifestations of *petit mal*, there are other equivalent manifestations, such as sudden jerkings in the limbs, sudden tremor, or a sudden visual sensation. Gowers gave no less than seventeen different manifestations. In occasional cases the patient has a sensation of losing his breath and may become red in the face.

After the attack the patient may be dazed for a few seconds and perform automatic actions, which may seem to be volitional. Undressing is common, but all sorts of odd actions may be performed, some of which are awkward or even serious. One patient after an attack was in the habit of tearing anything he could lay hands on, particularly books. Violent actions have been committed and assaults made, giving rise to questions which come before the courts. This condition has been termed masked epilepsy, or *epilepsia larvata*. In a majority of the cases of *petit mal* convulsions finally occur, at first slight, but ultimately major epilepsy is developed, and the attacks may then alternate.

(c) JACKSONIAN EPILEPSY.—This is also known as cortical, symptomatic, or partial epilepsy. It is distinguished from the ordinary epilepsy by the important fact that consciousness is retained or is lost late. The attacks are usually the result of irritative lesions in the motor zone, though there are probably also sensory equivalents of this motor form. Of 107 cases analyzed by Roland, there were 48 of tumor, 21 instances of inflammatory softening, 14 instances of acute and chronic meningitis, and 8 cases of trauma. The remaining instances were due to hæmorrhage or abscess, or were associated with sclerosis cerebri. Two other conditions may cause typical Jacksonian epilepsy—uræmia and general paresis. A considerable number of the cases of Jacksonian epilepsy are found in children following hemiplegia, the so-called post-hemiplegic epilepsy. The convulsions usually begin on the affected side, either in the arm or leg, and the fit may be unilateral and without loss of consciousness. Ultimately they become more severe and general.

In a typical attack the spasm begins in a limited muscle group of the face, arm, or leg. The zygomatic muscles, for instance, or the limb may twitch, or the toes may first be moved. Prior to the twitching the patient may feel a sensation of numbness or tingling in the part affected. The spasm extends and may involve the muscles of one limb only or of the face. The patient is conscious throughout and watches, often with interest, the march of the spasm. The onset may be slow, and there may be time for the patient to place a pillow on the floor, so as to be as comfortable as possible during the attack. The spasms may be localized for years, but there is a great risk that the partial epilepsy may become general.

(d) PYKNOLEPSY.—This is a form of minor epilepsy occurring between four and twelve years of age and usually with an explosive onset. The attacks are slight, of short duration, without any distress and may be many in a day. The child rarely falls, although the limbs relax, and objects are rarely dropped from the hands. The head may turn, the eyes rotate up and the arms show a feeble tonic spasm. There are no clonic spasms. The child

seems normal afterwards and the attacks are not harmful. Bromide and luminal apparently have no effect. The attacks stop spontaneously.

**Diagnosis.**—In *major epilepsy* the suddenness of the attack, the abrupt loss of consciousness, the order of the tonic and clonic spasm, and the relaxation of the sphincters are distinctive features. The convulsive seizures due to uræmia are usually readily recognized by the hypertension and the condition of the urine. Practically in young adults hysteria causes the greatest difficulty, and may closely simulate true epilepsy. Hysteroid attacks sometimes are post-epileptic. A careful study and observation of an attack usually make the diagnosis clear.

*Recurring epileptic seizures* in a person over thirty who has not had previous attacks is always suggestive of organic disease, usually syphilis.

*Petit mal* must be distinguished from attacks of syncope, and the vertigo of Ménière's disease, of a cardiac lesion, and of indigestion. Puzzling attacks occur especially about puberty, in which there is a slow and usually only partial loss of consciousness with dizziness, palpitation, slow pulse, low blood pressure and vaso-motor disturbance. The subjects have often grown rapidly. Diagnosis may be difficult if an attack is not observed. The results from good hygiene, supervised activity, and long hours of sleep are an aid.

*Jacksonian epilepsy* has features so distinctive and peculiar that it is at once recognized. It is, however, by no means easy always to determine upon what the spasm depends. Irritation in the motor centres may be due to a great variety of causes, among which tumors and localized meningo-encephalitis are the most frequent; but in uræmia localized attacks may occur. The most typical Jacksonian spasms are not infrequent in general paresis.

**Prognosis.**—This may be given to-day in the words of Hippocrates: "The prognosis in epilepsy is unfavorable when the disease is congenital, and when it endures to manhood, and when it occurs in a grown person without any previous cause. . . . The cure may be attempted in young persons, but not in old." Of cases beginning under ten years few are arrested, whereas of those beginning at puberty the opposite is true (W. A. Turner).

Death during the fit rarely occurs, but it may happen if the patient falls into water or a fire, or if the fit comes on while he is eating. Occasionally the fits stop spontaneously. This is particularly the case in the epilepsy in children which has followed the convulsions of teething or the fevers. Frequency of the attacks and marked mental disturbance are unfavorable indications. Some regard hereditary predisposition as favorable in the prognosis. The post-hemiplegic epilepsy is rarely arrested. Of seizures coming on in adults, those due to syphilis and local affections of the brain allow a more favorable prognosis.

**Treatment.**—GENERAL.—In the case of children the parents should be made to understand from the outset that epilepsy in the great majority of cases is an incurable affection, so that the disease may interfere as little as possible with the education of the child. The subjects need firm but kind treatment. Indulgence and yielding to caprices and whims are followed by weakening of the moral control, which is so necessary in these cases. Sources of irritation should receive attention. The disease does not incapacitate a person for all occupation. It is much better for an epileptic to have some definite pursuit but he should not follow an occupation which involves climb-

ing or working round machinery. He should not be allowed to drive a car or swim. The individual should take up an out-of-door occupation or have manual training suited to his condition. This is best done in an institution where he is carefully watched and studied. Psychoanalysis, with re-education, over a prolonged period is of value in some patients. There are many instances in which epileptics have been persons of extraordinary mental and bodily vigor, as, for example, Julius Caesar and Napoleon. One of the most distressing features is the mental impairment which follows in a certain number of cases. If such patients become extremely irritable or show signs of violence they should be placed under supervision in an institution. Marriage should be forbidden to epileptics.

During the attack a cork or bit of rubber should be placed between the teeth and the clothes should be loosened. The patient should be in the recumbent posture. If vomiting occurs he should be turned on the side and the mouth emptied of vomited material. As the attack usually passes off with rapidity, no special treatment is necessary, but in cases in which the convulsion is prolonged a few whiffs of chloroform or nitrite of amyl or a hypodermic of a quarter of a grain of morphia may be given.

DIETETIC.—The important points are to give the patient a light diet at fixed hours and on no account permit overloading of the stomach. Meat should not be given more than once a day. There are cases in which animal food seems injurious and a strict vegetable diet is sometimes useful. The possibility of protein sensitization should be considered and if found, these proteins should be excluded from the diet. The patient should not go to sleep until the completion of gastric digestion. The bowels should be kept freely open and colon irrigations are useful.

MEDICINAL.—The *bromides* are extensively used. They act as a depressant and therefore should be used only after a careful study of each patient. Sodium bromide is probably less irritating than the potassium salt and is better borne for a long period. It may be given in milk in which it is scarcely tasted. In all instances the dilution should be considerable. The dose for an adult should be from half a dram to a dram and a half (2 to 6 gm.) daily. The diet should be salt-free. It is often best to give but a single dose daily, about four to six hours before the attacks are most likely to occur. For instance, in nocturnal epilepsy 30 to 60 grains (2 to 4 gm.) should be given in the evening. If the attack occurs in the morning, the patient should take a full dose when he awakes. When given three times a day it is less disturbing after meals. Each patient should be carefully studied to determine how much bromide should be used. The individual susceptibility varies and some patients require more than others. Fortunately, children take the drug well and stand proportionately larger doses than adults. Saturation is indicated by certain unpleasant effects, particularly drowsiness, mental torpor, and gastric and cardiac distress. Loss of palate reflex is one of the earliest indications. A common feature is the development of acne, which is no indication of bromism. The tendency to this is diminished by giving the drug largely diluted in alkaline waters and administering arsenic from time to time. Written directions should be given to the mother or the friends of the patient, and he should not be held responsible for the administration of the medicine. The addition of belladonna to the bromide is recommended.

*Pheno-barbital* (luminal) is probably more useful than bromide, beginning with doses of one grain (0.065 gm.) one to three times a day and gradually increasing until the best dosage is found. Amounts over six grains a day should be given only for short periods. The drug may be given for four or five days a week and bromide given on the other days. If attacks occur at certain times, e.g., at menstruation, the drugs should be given in larger doses for a few days before. In status epilepticus, hyoscine or morphine hypodermically may be useful.

Among other remedies recommended are chloral, cannabis indica, and nitroglycerin. Nitroglycerin is sometimes advantageous in *petit mal* and must be given in full doses, from 2 to 5 drops of the 1 per cent. solution, and increased until the physiological effects are produced. Calcium lactate in 20 grain (1.3 gm.) doses daily has been recommended. Counter-irritation is rarely advisable. When the aura is very definite and constant in its onset, as from the hand or from the toe, a ligature tightly applied may stop the oncoming fit.

The subjects of a chronic and, in most cases, an incurable disease, epileptic patients form no small portion of the unfortunate victims of charlatans and quacks, who prescribe to-day, as in the time of the father of medicine, "purifications and spells and other illiberal practices of like kind."

**SURGICAL.**—In Jacksonian epilepsy the propriety of surgical interference is universally granted. It is questionable whether in the epilepsy following hemiplegia it is likely to be of any benefit. In idiopathic epilepsy, when the fit starts in a certain region—the thumb, for instance—and the signal symptom is invariable, the centre controlling this part may be removed. Operation, *per se*, appears in some cases to have a curative effect. The operation in the traumatic epilepsy, as after fracture, is much more hopeful. Operations have not been always on the skull, and White collected an interesting series in which various surgical procedures had been done, often with curative effect, such as ligation of the carotid artery, castration, excision of the superior cervical ganglia, incision of the scalp, etc.

## VI. MIGRAINE

(*Hemicrania; Sick Headache*)

**Definition.**—A paroxysmal affection characterized by severe headache, usually unilateral, and often associated with vomiting or disorders of vision.

**Etiology.**—Heredity plays an important rôle in 90 per cent. of cases according to Möbius. Women and members of neurotic families are most frequently attacked. Many distinguished men have been its victims, and the astronomer Airy gave a classical account of his case. The nature of the disease is in dispute, and many views have been entertained:

(a) That it is a *toxæmia* from disorder of the intestinal digestion or from some self-manufactured poison.

(b) That it is a *vasomotor* affection with spasm of the arteries, in favor of which are the facts that in the attack the temporal arteries on the affected side may be felt to be small, the retinal arteries may sometimes be seen in

spasm, and sclerosis of the arteries on the same side is found in a certain number of cases. A striking confirmation is the temporary paralysis which may be associated with an attack of monoplegic or hemiplegic character. Mitchell Clarke reported a history of recurring motor paralysis in eleven members in three generations of the same family. The characteristic visual phenomena preceded the unilateral headache, especially the hemiopia. In most of the attacks the hemiplegia was on the right side. It lasted from a few hours to a day and disappeared completely. It is difficult to explain such cases except on the view of a transient spasm of the arteries.

(c) Others regard the affection as of *reflex* origin arising from a refractive error in the eyes, or from trouble in the nose or sexual organs.

(d) A constitutional anatomical defect causing stenosis of the foramen of Monro, associated with hyperæmia of the choroid plexus and increase of pressure in one or both lateral ventricles, is the view of Spitzner.

(e) Some regard it as a disturbance of the sympathetic nervous system, which may be caused by a number of conditions. The variety of causal factors is a striking feature of the disease.

The majority of cases begin in young adults, but Sinclair refers to a case in a child of two years. Many circumstances bring on the attack: a powerful emotion of any sort, mental or bodily fatigue, digestive disturbances, or the eating of some particular food. The subjects often suffer from train or sea sickness. The paroxysmal character is one of the most striking features of the attacks which may occur on the same day every week, every fortnight, or every month. Headaches of the migraine type may occur for years with chronic nephritis and with tumors and other lesions of the base of the brain.

**Symptoms.**—Premonitory signs are present in many cases, and the patient can tell when an attack is coming on. Remarkable prodromata have been described, particularly in connection with vision. Apparitions may appear—visions of animals, such as mice, dogs, etc. Transient hemianopia or scotoma may be present. In other instances there is spasmodic action of the pupil on the affected side, which dilates and contracts alternately, the condition known as *hippus*. Frequently the disturbance of vision is only a blurring or there are balls of light, or zigzag lines, or the so-called fortification spectra (teichopsia), which may be illuminated with gorgeous colors. Disturbances of the other senses are rare. Numbness of the tongue and face and occasionally of the hand may occur with tingling. More rarely there are cramps or spasms in the muscles of the affected side. Transient aphasia may occur and be intermittent. The *paralysis* may be (1) of cerebral origin—hemiplegia or aphasia, or (2) due to lesions of cranial nerves—optic nerve and ophthalmoplegias, the oculomotor most often, abducens rarely, trochlearis very rarely. The supposed involvement of the facial is relapsing facial palsy in migraine (Ramsay Hunt). Some patients show marked *psychical* disturbance, either excitement or, more commonly, mental confusion or great depression. Dizziness occurs in some cases. The *headache* follows a short time after the prodromal symptoms or may be the feature of onset, especially on waking in the morning. It is cumulative and expansile in character, beginning as a localized small spot, which is generally constant either on the temple or forehead or in the eyeball. It is usually described as of a penetrating, sharp,

boring character. The pain gradually spreads and involves the entire side of the head, sometimes the neck, and may pass into the arm. In some cases both sides are affected. *Nausea* and *vomiting* are common and, if the attack comes on when the stomach is full, vomiting may give relief. *Vasomotor* symptoms may be present. The face may be pale, and there may be a marked difference between the two sides. Subsequently the face and ear on the affected side may become a burning red from the vasodilator influences. The pulse may be slow. The temporal artery on the affected side may be firm and hard, and arterio-sclerotic—confirmed anatomically by Thoma. Few affections are more prostrating and during the paroxysm the patient may scarcely be able to raise the head from the pillow. The slightest noise or light aggravates the condition.

The *duration* of the attack is variable. The severer forms usually incapacitate the patient for two or three days. In other instances the entire attack is over in a day. The disease recurs for years, and in patients with a marked hereditary tendency may persist throughout life. In women the attacks often cease after the menopause, and in men after the age of fifty.

**Diagnosis.**—In well-marked cases there is little difficulty, especially with a clear history, but nephritis and intracranial tumor should be considered in cases originating in later life. There are abortive attacks, in which a great variety of symptoms occur, not easily recognized until perhaps a typical attack appears.

**Treatment.**—The patient is usually aware of the causes which precipitate an attack. Avoidance of excitement, regularity in the meals, and moderation in diet are important rules. Some patients suffer from too much protein and others from too much carbohydrate. It is well to try a low carbohydrate diet and if no benefit results cut out animal protein for a time. Some patients are helped by exclusion of eggs from the diet, others by a strict vegetarian diet. The intravenous injection of peptone has helped some patients. Treatment should be directed toward the removal of the conditions upon which the attacks depend. Much may be done by watchfulness and care in regulating the bowels and watching the diet. Errors of refraction should be adjusted. On no account should children with migraine be allowed to compete in school for prizes. A prolonged course of bromides sometimes proves successful. If anæmia is present, iron and arsenic should be given. When the arterial tension is increased a course of nitroglycerin may be tried. Not too much, however, should be expected from preventive treatment as in a large proportion of cases the headaches recur in spite of all we (including the refractionists) can do. Lavage of the stomach with water at 105°, a brisk saline cathartic and irrigation of the colon with hot saline solution are sometimes of value at the onset. Alkaline water should be taken freely by mouth. During the paroxysm the patient should be kept in bed and absolutely quiet. If the patient feels faint and nauseated a small cup of strong coffee may give relief. A prolonged course of *cannabis indica* may be tried. Antipyrine and phenacetine have been much used. When given early, at the very outset, they are sometimes effective. Small, repeated doses are more satisfactory. Lauder Brunton advised a combination of sodium salicylate (gr. 15, 1 gm.) and potassium bromide (gr. 30, 2 gm.) at the onset. Combinations of acetyl-salicylic acid, caffeine and phenacetine may be used.

Pheno-barbital (gr.  $\frac{1}{2}$  to 0.03 to 0.06 gm.) given daily for a considerable period is sometimes useful. Calcium lactate (gr. 30, 2 gm.) at the onset may avert an attack. Of other remedies, nux vomica, gelsemium, and ergot have been recommended.

**Ophthalmoplegic Migraine.**—This term was applied by Charcot to a special form in which there is weakness or paralysis of one or more eye muscles, with or after a migraine attack. The oculo-motor nerve is usually involved. Ptosis, loss of certain movements, and double vision are the common features, which may persist for some days. Local causes, especially syphilis, should be excluded before the diagnosis is established.

## VII. NEURALGIA

**Definition.**—A painful affection of the nerves, due to functional disturbance of their central or peripheral extremities or to neuritis in their course.

**Etiology.**—Members of neuropathic families are most subject to the disease. It affects women more than men. Children are rarely attacked. Of all causes debility is the most frequent. It is often the first indication of an enfeebled nervous system. The various forms of anemia are frequently associated with neuralgia. It may be a prominent feature at the onset of certain acute diseases, particularly typhoid fever. It has not been shown that neuralgia is more frequent in malarial districts, but it occasionally occurs in malarial cachexia. Exposure to cold is a cause in susceptible persons. Reflex irritation, particularly from carious teeth, and disease of the antrum and frontal sinuses are common causes of neuralgia of the fifth nerve. The disease occurs sometimes in gout, lead poisoning, and diabetes. Persistent neuralgia may be a feature of latent nephritis.

**Symptoms.**—Before the onset of the pain there may be uneasy sensations sometimes tingling in the part which will be affected. The pain is localized to a certain group or division of nerves, usually affecting one side. The pain is not constant, but paroxysmal, and is described as stabbing, burning, or darting in character. The skin may be exquisitely tender in the affected region, particularly over certain points along the course of the nerve, the so-called tender points. Movements, as a rule, are painful. Trophic and vaso-motor changes may accompany the paroxysm; the skin may be cool, and subsequently hot and burning; occasionally local edema or erythema occurs. More remarkable still are the changes in the hair, which may become blanched (canities) or fall out. Fortunately, such alterations are rare. Twitchings of the muscles, or even spasms, may be present during the paroxysm. After lasting a variable time—from a few minutes to many hours—the attack subsides. Recurrence may be at definite intervals—every day at the same hour, or at intervals of two, three, or even seven days. Occasionally the paroxysms develop only at the catamenia.

### CLINICAL VARIETIES, DEPENDING ON THE NERVE ROOTS AFFECTED

**Trigeminal Neuralgia; Tic Douloureux.**—A distinction must be drawn between the minor and major neuralgias of the fifth cranial nerve. The former

may be symptomatic of the involvement of the ganglion or one of its branches in some disease process—the pressure of a tumor on the ganglion, carious teeth, sinus infection or injury. There may be referred pains in this area from morbid processes within the cranium, or from visceral disease elsewhere. A painful neuralgia may follow an attack of zoster in any division of the fifth nerve.

The *major trigeminal neuralgia* is a primary affection of the Gasserian ganglion. The designation *tic* is not descriptive; there is usually immobility. The sex incidence is about equal; the majority of cases begin between the ages of forty and sixty. No definite etiological factor is evident. The right side is involved in about two-thirds of the cases. Patrick's figures show that the second and the third branches are involved more often than the first. It begins most often in the second branch and later two or all three branches may be involved. The *pain* is of sudden onset, usually excruciating and in paroxysms, which may recur, usually not lasting longer than two minutes. The attacks are excited by any external irritation which may be very slight, such as a draught of air, touching the skin, and the movements in speaking, eating or swallowing. The areas over which irritation excites the pain are termed *doloro-genetic* or "trigger" zones. These do not always correspond to the pain zone. The pain may radiate into the cervical nerves or down the arms. The attacks tend to be of increasing severity and in advanced cases the paroxysms may recur at short intervals in steady succession.

The *diagnosis* is rarely in doubt but minor forms should not be mistaken for the major. The pain in them is paroxysmal, so that a steady pain about the face is probably not trifacial neuralgia. If the area has been rubbed or massaged, or the patient touches it to show where the pain is, the disease is probably not the major form. The disease may be remittent but tends to progress and increase in severity so that life is almost unbearable. There are certain other forms of facial neuralgia in addition to the trigeminal ones noted: (1) Attributed to the spheno-palatine (Meckel's) ganglion (Sluder), in which an inflammatory process in the sinuses spreads to the ganglion. There is pain about the eye, root of the nose, and upper jaw back to the mastoid which may spread more widely. The pain is constant or paroxysmal. (2) Genuiculate ganglion. Hunt has emphasized the involvement of the sensory part of the facial nerve. The auricular herpetic eruption is due to disease of the geniculate ganglion. (3) A painful convulsive tic occurs; and (4) there is an intractable form in run-down nervous patients, for which no definite cause can be found.

**Cervico-occipital neuralgia** involves the posterior branches of the first four cervical nerves, particularly the inferior occipital, at the emergence of which there is a painful point about half-way between the mastoid process and the first cervical vertebra. It may be caused by cold or be due to cervical caries. Surgical measures may be required if the pain is severe.

**Cervico-brachial neuralgia** involves the sensory nerves of the brachial plexus, particularly in the cubital division. When the circumflex nerve is involved the pain is in the deltoid. The pain is most commonly about the shoulder and down the course of the ulnar nerve. There is usually a marked tender point upon this nerve at the elbow. This form rarely follows cold, but more frequently results from arthritis and trauma.

**Neuralgia of the phrenic nerve** is rare. It is sometimes found in pleurisy and in pericarditis. The pain is chiefly at the lower part of the thorax on a line with the insertion of the diaphragm, and here may be painful points on deep pressure. Full inspiration is painful, and there is great sensitiveness on coughing or any movement by which the diaphragm is suddenly depressed.

**Intercostal Neuralgia.**—This is most frequent in women and common in hysteria. Post-zoster neuralgias are common in this situation. The possibility of spinal disease, of tumor, spondylitis, caries, or aneurism must be borne in mind.

**Lumbar Neuralgia.**—The posterior fibres of the lumbar plexus, particularly the ilio-scrotal branch, are affected. The pain is in the region of the iliac crest, along the inguinal canal, in the spermatic cord, and in the scrotum or labium majus. The affection known as irritable testis, probably a neuralgia of this nerve, may be severe and accompanied by syncopal sensations.

**Coccydynia.**—This is regarded as a neuralgia of the coccygeal plexus. It is most common in women, and is aggravated by the sitting posture. It is very intractable, and may necessitate the removal of the coccyx, an operation, however, which is not always successful.

**Neuralgias of the Nerves of the Feet.**—Many of these cases accompany varying degrees of flat-foot. The condition is brought about by weakness or fatigue of the muscles supporting the arches of the foot, which consequently settle until the strain of the superimposed body-weight falls upon the ligamentous and aponeurotic attachments between the metatarsal and tarsal bones. Rest, massage, exercises, and orthopaedic measures are indicated.

**PAINFUL HEEL.**—Both in women and men there may be about the heel severe pains which interfere seriously with walking—the pododynia of S. D. Gross. There may be little or no swelling, no discoloration, and no arthritis. Some cases follow a gonococcus infection and are due to a bony spur.

**PLANTAR NEURALGIA.**—This is often associated with a definite neuritis, such as follows typhoid fever, and has been seen in an aggravated form in caisson disease (Hughes). The pain may be limited to the tips of the toes or to the ball of the great toe. Numbness, tingling, and hyperæsthesia or sweating may occur with it.

**METATARSALGIA.**—Thomas G. Morton's "painful affection of the fourth metatarso-phalangeal articulation" is a peculiar and very trying disorder, seen most frequently in women, and usually in one foot. Morton regarded it as due to a pinching of the metatarsal nerve. It usually requires operation. In some cases there is a *metatarso-phalangeal osteochondritis*. Lewis (1923) found 65 cases reported. Trauma and infection are factors. There is pain referred to the head of the metatarsal bone, with swelling, due to exudate, and tenderness. The X-ray study is diagnostic. In treatment, weight should be taken entirely off the foot, which is placed in a cast.

**CAUSALGIA (Thermalgia).**—A form of neuralgia following gunshot wounds, most frequently of the median and of the sciatic branches, characterized by burning throbbing pains of the greatest intensity, glossy skin, vasomotor disturbances, and at last a condition of general hyperæsthesia and nervousness that makes life unbearable. Nothing has been added to Weir Mitchell's classical description (1864), and later he gave the above name from the

Greek words for burning and pain. Many cases were seen in the late war, and Stopford suggested the name *thermalgia*. An explanation of *causalgia* is difficult. The median and post-tibial nerves have a large number of vasomotor fibres, interference with which may cause the peculiar character of the pain; indeed, it has been suggested that the pain is caused by irritation of the peri-arterial sympathetic fibres and not by the wound of the nerve itself. Anatomically, partial division with perineural and intra-neural fibrosis is present, but these are found in scores of cases in which *causalgia* is not present.

**VISCERAL NEURALGIAS.**—The more important of these occur with cardiac and gastric neuroses. They are most frequent in women often with *neurasthenia* and *hysteria*. The referred pains represent a "viscero-sensory reflex." With lesions in the viscera there is surface tenderness brought out by pinching the skin or applying a sharp object. The distribution of the sensory nerves and their segmental relations are important in working out the source of the disturbance.

**Treatment of Neuralgia.**—Causes of reflex irritation should be remedied and foci of infection removed. The neuralgia, as a rule, recurs unless the general health improves; so that tonic and hygienic measures of all sorts should be employed. Often a change of air or surroundings relieves a severe neuralgia. Obstinate cases may be cured by a prolonged residence in the mountains, with an out-of-door life and plenty of exercise. A strict vegetable diet will sometimes relieve the neuralgia or headache of a gouty person. Of general remedies, iron is often a specific in the cases associated with *anæmia*. Arsenic is beneficial in these forms, and should be given in ascending doses. The value of quinine has been much overrated. It probably has no more influence than any other bitter tonic, except in the rare instances in which the neuralgia is definitely associated with *malaria*. Strychnine, cod-liver oil, and phosphorus are advantageous. Of remedies for the pain, *phenacetine* and *acetyl-salicylic acid* are generally useful, and small doses of *codeine* may be added if necessary. *Morphia* should not be given. *Gelsemium* is highly recommended. *Valerian* and ether, which often act well together, may be given. In the minor form of *trigeminal neuralgia* *nitro-glycerine* in large doses may be tried. Dana has seen good results follow rest with large doses of *strychnia* given hypodermically. *Aconitin* in doses of one two-hundredth of a grain (0.00032 gm.) may be tried. *Diathermy* may be useful.

Of local applications, the cautery is invaluable, particularly in the more chronic forms. *Acupuncture* may be used. Counter-irritation and the use of heat and various forms of light therapy are helpful. *Chloroform liniment*, *camphor* and *chloral*, and *menthol*, may be tried. Freezing over the tender point with ether spray is sometimes successful. The continuous current may be used. The sponges should be warm, and the positive pole should be placed near the seat of the pain. The current should cause a slight tingling or burning, but not pain.

For *trigeminal neuralgia* there are two successful measures, (1) injection of alcohol into the branch, the trunk or the ganglion itself, often satisfactory in skilled hands; and (2) removal of the ganglion. Cushing's results show the remarkable benefit which may result.

### VIII. PROFESSIONAL SPASMS; OCCUPATION NEUROSES

The continuous and excessive use of the muscles in performing a certain movement may be followed by an irregular, involuntary spasm or cramp, which may completely check the performance of the action. The condition is found frequently in writers, hence the term *writer's cramp* or *scrivener's palsy*, and also occurs in typists, needle-workers, piano and violin players, telegraph operators, milkmaids, weavers, and cigar and cigarette rollers.

The most common form is *writer's cramp*, which is much more frequent in men than in women. Of 75 cases reported by Poore, all the instances of undoubted writer's cramp were in men. An investigation by Thompson and Sinclair into telegraphist cramp in England shows that the disease is rare, only 13 cases among 7,000-8,000 employees. Persons of a nervous temperament are more liable to the disease. Occasionally it follows slight injury. In a majority a faulty method of writing has been employed, using either the little finger or the wrist as the fixed point. Persons who write with the middle of the forearm or the elbow as the fixed point are rarely affected.

No anatomical changes have been found. The most reasonable explanation of the disease is that it results from a deranged action of the nerve centres presiding over the muscular movements involved in the act of writing, a condition which has been termed *irritable weakness*.

**Symptoms.**—These may be described under five heads (Lewis).

(a) **CRAMP OR SPASM.**—This is often an early symptom and most commonly affects the forefinger and thumb; or there may be a combined movement of flexion and adduction of the thumb, so that the pen may be twisted from the grasp and thrown to some distance. Weir Mitchell described a lock-spasm, in which the fingers become so firmly contracted upon the pen that it can not be removed.

(b) **PARESIS AND PARALYSIS.**—This may occur with the spasm or alone. The patient feels a sense of weakness and debility in the muscles of the hand and arm and holds the pen feebly. Yet the grasp of the hand may be strong and there may be no paralysis for ordinary acts.

(c) **TREMOR.**—This is most commonly seen in the forefinger and may be a premonitory symptom of atrophy. It is not an important symptom, and is rarely sufficient to produce disability.

(d) **PAIN.**—Abnormal sensations, particularly a tired feeling in the muscles, are very constantly present. Actual pain is rare, but there may be irregular shooting pains in the arm. Numbness or soreness may exist. If, as sometimes happens, a subacute neuritis develops, there may be pain over the nerves and numbness or tingling in the fingers.

(e) **VASO-MOTOR DISTURBANCES.**—These may occur in severe cases. There may be hyperæsthesia. Occasionally the skin becomes glossy, or there is a condition of local asphyxia resembling chilblains. In attempting to write, the hand and arm may become flushed and the veins increased in size. Early the electrical reactions are normal, but in advanced cases there may be diminution of faradic and sometimes increase in the galvanic irritability.

**Diagnosis.**—A well-marked case of writer's cramp or palsy could scarcely be mistaken. Care must be taken to exclude the existence of any cerebral

spinal disease, such as progressive muscular atrophy or hemiplegia, or local affection, such as cervical rib. The physician is sometimes consulted by nervous persons who fancy they are becoming subject to the disease and complain of stiffness or weakness without displaying any characteristic features.

**Prognosis.**—The course is usually chronic. If taken in time and if the hand is allowed perfect rest, the condition may improve rapidly, but too often there is a strong tendency to recurrence. The patient may learn to write with the left hand, but this also may after a time be attacked.

**Treatment.**—Various prophylactic measures have been advised. It is important that a proper method of writing be adopted. Gowers suggested that if all persons wrote from the shoulder writer's cramp would practically not occur. Various devices have been invented for relieving the fatigue, but none of them are very satisfactory. The use of the type-writer has diminished the frequency of scrivener's palsy. Rest is essential and no measures are of value without it. Massage and manipulation, when combined with systematic gymnastics, give the best results. The patient should systematically practise the opposite movements to those concerned in the cramp. This muscle training often gives good results. Poore recommends the galvanic current applied to the muscles, which are at the same time rhythmically exercised. In very obstinate cases the condition remains incurable.

## IX. HYSTERIA

**Definition.**—A disorder of personality manifested by a heightened and perverted suggestibility, a change in character, together with certain mental and bodily states induced by suggestion—auto or hetero—and cured by persuasion. It is "an irrational answer to a conflict."

**Etiology.**—Persons with mobile emotional dispositions, especially women, are the chief subjects. In periods of great stress, as in the recent war, it becomes a widespread and serious disorder. A community disease, often spreading widely in institutions, such as schools and convents, it may behave like an epidemic, as in the dancing mania. The essential element under the above definition is the first—*heightened suggestibility*. (a) With the chameleon we take the color of our surroundings. The company, physical conditions, the weather, etc., send our spirits up and down like the mercury in a barometer. Suggestion, deliberate by speech, unconscious through imitation, is the most important part of education, and to free the mind as far as possible from the mastery of these external influences has been the goal from the days of the Greeks. Love, hate, and fear, the three powerful emotions, control us individually or sway us in herds as the cattle on the plains. The dominant influence of suggestion is everywhere in the story of human progress; just as it is in the black chapters of superstition, folly, and crime. Unconscious imitation, or an imitation against which the individual is powerless to fight, has been the important factor in outbreaks of hysteria as the dancing mania, the epidemic chorea, and such tragedies as led to the persecutions for witchcraft.

(b) Right judgments are indispensable conditions to right action in mind or muscle and it is in this Stoic doctrine of the control of the will—

the will to do and the will to avoid—that we find the key to many of the problems of hysteria. It may be a knee “locked” for months. An injury or pain induces the fixed belief that the joint can not be moved, loss of muscle judgment—there were scores of such war cases—but ten minutes at Seale-Hayne or a trip to Lourdes and the joint is flexible. After the shock of an explosion a man is blind (without a lesion), the condition persists—the judgment has been lost—to be restored months afterwards at a temple of Esculapius or by some modern Galen. An emotional girl takes an aversion to her mistress. The moral judgment is lost and she begins to play pranks, sometimes harmless, but often serious as entailing great inconvenience and loss, as in the Norfolk case in which the walls of a house were so covered with paraffin, sandal oil and water that it had to be abandoned. Or craving sympathy, she will inflict all sorts of injuries, even wound herself to such an extent as to necessitate amputation of a limb. Loss of right judgment then in muscle action, sense action, and conduct are essential factors. As the impulse—suggestion—is spontaneous we speak of it as auto-suggestion—and in direct proportion to the feebleness of control by the will is the readiness with which muscles, sense and mind yield to impulses not prompted by right judgment.

Charcot and his followers regarded hysteria as a psychosis, in which morbid states are induced by ideas. The capability of responding to suggestion is the test of its existence. It is a disturbance in the sphere of personality, in which the emotions have an exaggerated influence on the sensory, motor and secretory functions. Babinski holds that hysteria is a mental condition with certain primary phenomena and certain secondary accidental symptoms. The essence of the primary features is that they may be produced by suggestion, and may be made to disappear by persuasion (pithiatism). The primary symptoms include hemianæsthesia, paralysis, contractures, etc.; secondary features, as muscular atrophy, are directly dependent upon the primary and can not themselves be induced by suggestion.

In the Breuer-Freud theory we return to the days of Aretæus, who originated (?) the views of sexual hysteria and believed the womb, “like an animal within an animal” and altogether erratic, caused all sorts of trouble in its wanderings. Freud’s view is thus analyzed by Jelliffe in his article in our *System of Medicine* (2nd Ed., Vol. V.). “There develop usually on a constitutional basis, in the period before puberty, definite sexual activities which are mostly of a perverse nature. These activities do not, as a rule, lead to a definite neurosis up to the time of puberty, which in the psychic sphere appears much earlier than in the body, but sexual phantasy maintains a perverse constellated direction by reason of the infantile sexual activities. On constitutional (affect) grounds the increased fantasy of the hysteric leads to the formation of complexes which are not taken up by the personality and by reason of shame or disgust remain buried. There, therefore, results a conflict between the characteristic normal libido and the sexual repressions of these buried infantile perversions. These conflicts give rise to the hysterical symptoms. It is in his contributions to the sexual theory that Freud develops his later thoughts of the sexual origin of the hysterical reaction. By sexual it is important to remember that Freud is not speaking of sensual.

The significance of Freud's theory is the tracing of every case to sexual traumas during early childhood. Sexual experiences differ, however, from ordinary experiences—the latter have a tendency to fade out, while the idea of the former grows with increasing sexual maturity. There results a disproportionate capacity for increased reaction which takes place in the subconscious. This is the cause of the mischief.

"There must be, however, a connecting link between the infantile sexual traumata and the later manifestations. This connection Freud finds in the so-called 'hysterical fancies.' These are the day-dreams of erotic coloring, wish-gratifications, originating in privation and longing. These fancies hark back to the original traumatic moment, and, either originating in the subconscious or shortly becoming conscious, are transformed into hysterical symptoms. They constitute a defence of the ego against the revival, as reminiscences, of the repressed traumatic experiences of childhood" (White).

The affection is most common in females, and usually appears first about the time of puberty, but the manifestations may continue until the menopause, or even until old age. Men are by no means exempt, and hysteria in the male is not rare. It occurs in all races, but is much more prevalent, particularly in its severer forms, in members of the Latin race. In England and the United States the milder grades are common, but the graver forms are rare in comparison with the frequency with which they are seen in France.

Children under twelve years of age are not very often affected, but the disease may be well marked as early as the fifth or sixth year. One of the saddest chapters in the history of human deception, that of the Salem witches, might be headed *hysteria in children*, since the tragedy resulted directly from the hysterical pranks of girls under twelve years of age.

Of predisposing causes, two are important—*heredity* and *education*. The former acts by endowing the child with a mobile, abnormally sensitive nervous organization. We see cases most frequently in families with marked neuro-pathic tendencies, the members of which have suffered from neuroses of various sorts. Education at home too often fails to inculcate habits of self-control. A child grows to girlhood with an entirely erroneous idea of her relations to others, and accustomed to have every whim gratified and abundant sympathy lavished on every woe, however trifling; she reaches womanhood with a moral organization unfitted to withstand the cares and worries of every-day life. At school, between the ages of twelve and fifteen, when the vital energies are absorbed in the rapid development of the body, she is often cooped in close school rooms for six or eight hours daily. The result too frequently is an active, bright mind in an enfeebled body, ill adapted to subserve the functions for which it was framed, easily disordered, and prone to react abnormally to the ordinary stimuli of life. Among the more direct influences are emotions of various kinds, fright occasionally, more frequently love affairs, grief, and domestic worries. Physical causes less often bring on hysterical outbreaks, but they may follow an injury or develop during the convalescence from an acute illness or be associated with disease of the generative organs.

"**Chorea major**": "**Pandemic Chorea**."—The common name, St. Vitus's dance, applied to chorea has come to us from the Middle Ages, when under the influence of religious fervor there were epidemics characterized by great excitement, gesticulations, and dancing. For the relief of these symptoms, when

excessive, pilgrimages were made, and, in the Rhenish provinces, particularly to the Chapel of St. Vitus in Zebern. Epidemics of this sort occurred also during the nineteenth century, and descriptions of them among the early settlers in Kentucky have been given by Robertson and Yandell. It was unfortunate that Sydenham applied the term chorea to an affection in children totally distinct from this chorea major, which is in reality an hysterical manifestation under the influence of religious excitement.

**Symptoms.**—A useful division is into the convulsive and non-convulsive varieties.

**CONVULSIVE HYSTERIA.**—(a) *Minor Forms.*—The attack, commonly following emotional disturbance, sets in suddenly or may be preceded by symptoms, called by the laity "hysterical," such as laughing and crying alternately, or a sensation of constriction in the neck, or of a ball rising in the throat—the *globus hystericus*. Sometimes, preceding the convulsive movements, there may be painful sensations arising from the pelvic, abdominal, or thoracic regions. From the description these sensations resemble auræ. They become more intense with the rising sensation of choking in the neck and difficulty in getting breath, and the patient falls into a more or less violent convulsion. The fall is not sudden, as in epilepsy, but the subject goes down, as a rule, easily, often picking a soft spot, like a sofa or an easy-chair, and in the movements apparently exercises care to do herself no injury. Yet at the same time she appears to be unconscious. The movements are clonic and disorderly, with the head and arms thrown about in an irregular manner. The paroxysm after a few minutes slowly subsides, then the patient becomes emotional, and gradually regains consciousness. When questioned the patient may confess to having some knowledge of the events which have taken place, but, as a rule, has no accurate recollection. During the attack the abdomen may be much distended with flatus, and subsequently a large amount of clear urine may be passed. These attacks vary greatly; there may be scarcely any movements of the limbs, but after a nerve storm the patient sinks into a torpid, semi-unconscious condition, from which she is roused with difficulty. In some cases the patient passes from this state into a condition of catalepsy.

(b) *Major Forms; Hystero-epilepsy.*—Typical instances are very rare in the United States and in England. The attack is initiated by certain prodromata, chiefly minor hysterical manifestations, either foolish or unseemly behavior, excitement, sometimes dyspeptic symptoms with tympanites, or frequent micturition. Areas of hyperæsthesia may be marked, the so-called hysterogenic spots described by Richet. These are usually symmetrical and situated over the upper dorsal vertebra, and in front in a series of symmetrically placed areas on the chest and abdomen. Painful sensations or a feeling of oppression and a *globus* rising in the throat may be complained of prior to the onset of the convulsion, which, according to French writers, has four distinct stages: (1) Epileptoid condition, which closely simulates a true epileptic attack with tonic spasm (often leading to opisthotonos), grinding of the teeth, congestion of the face, followed by clonic convulsions, gradual relaxation, and coma. (2) Succeeding this is the period which Charcot has termed *clownism*, in which there is an emotional display and a remarkable series of contortions or of cataleptic poses. (3) Then in typical cases there is a stage in which the patient assumes certain attitudes expressive of ecstasy, fear, beatitude, or

eroticism. (4) Finally consciousness returns and the patient enters upon a stage in which she may display very varied symptoms, chiefly manifestations of a delirium with extraordinary hallucinations. Visions are seen, voices heard, and conversations held with imaginary persons. In this stage patients will relate with the utmost solemnity imaginary events, and make extraordinary and serious charges against individuals. This sometimes gives a grave aspect to these seizures, for not only does the patient make and believe the statements, but when recovery is complete the hallucination sometimes persists. After an attack a patient may remain for days in a state of lethargy or trance.

**NON-CONVULSIVE FORMS.**—So complex and varied is the picture that the manifestations are best considered according to the systems involved.

(a) *Disorders of Motion.*—(1) *Paralysis.*—These may be hemiplegic, paraplegic, or monoplegic. Hysterical diplegia is extremely rare. The paralysis either sets in abruptly or gradually, and may take weeks to attain its full development. *There is no type or form of organic paralysis which may not be simulated in hysteria.* Sensation is either lessened or lost on the affected side. The hysterical paraplegia is more common than hemiplegia. The loss of power is not absolute; the legs can usually be moved, but do not support the patient. The reflexes may be increased, though the knee-jerk is often normal. A spurious ankle clonus may sometimes be present. The feet are usually extended and turned inward in the equino-varus position. The muscles do not waste and the electrical reactions are normal. Other manifestations, such as paralysis of the bladder or aphonia, are usually associated. Hysterical monoplegias may be facial, crural, or brachial. A condition of ataxia sometimes occurs with paresis. Inco-ordination may be a marked feature, and there are usually sensory manifestations.

The following points are important in deciding between functional and organic hemiplegia. The absence of epigastric and cremasteric reflexes with Babinski's sign suggests organic disease. If the patient lies on a table, with the arms folded on the chest and the legs apart, and is asked to sit up without using the arms, in doing this the organically paralyzed leg is flexed at the hip and the heel raised from the table. The heel of the non-paralyzed leg is pressed against the table. In hysterical paralysis both heels remain pressed against the table. Another test is made with the patient lying on the back. When asked to raise the unaffected leg, the opposite leg, paralyzed for voluntary effort, is strongly pressed down (Hoover).

(2) *Contractures and Spasms.*—The hysterical contractures may attack almost any group of voluntary muscles and be of the hemiplegic, paraplegic, or monoplegic type. They may come on suddenly or slowly, persist for months or years, and disappear rapidly. The contracture is most common in the arm, which is flexed at the elbow and wrist, while the fingers tightly grasp the thumb in the palm of the hand; more rarely the terminal phalanges are hyperextended. It may occur in one or in both legs, more commonly in one. Ankle clonus is present; the foot is inverted and the toes are strongly flexed. These cases may be mistaken for lateral sclerosis and the difficulty in diagnosis may be very great. The spastic gait is typical, and with the exaggerated knee-jerk and ankle clonus the picture may be characteristic. Other forms of contracture may be in the muscles of the hip, shoulder, or neck;

more rarely in those of the jaws—hysterical trismus—or in the tongue. There are remarkable local contractures in the diaphragm and abdominal muscles, producing a phantom tumor, in which just below and in the neighborhood of the umbilicus is a firm, apparently solid growth. According to Gowers, this is produced by relaxation of the recti and a spasmodic contraction of the diaphragm, together with inflation of the intestines with gas and an arching forward of the vertebral column. They are apt to occur in middle-aged women about the menopause, and are frequently associated with symptoms of spurious pregnancy—*pseudo-cyesis*. The resemblance to a tumor may be striking. The only safeguard is in complete anæsthesia, when the tumor entirely disappears. Mitchell reported an instance of a phantom tumor in the left pectoral region just above the breast, which was tender, hard, and dense.

*Rhythmic Hysterical Spasm.*—The movements may be of the arm, either flexion and extension, or, more rarely, pronation and supination. Clonic contractions of the sterno-cleido-mastoid or of the muscles of the jaws or of the rotatory muscles of the head may produce rhythmic movements of these parts. The spasm may be in one or both psoas muscles, lifting the leg in a rhythmic manner eight or ten times in a minute. In other instances the muscles of the trunk are affected, and every few moments there is a bowing movement—salaam convulsions—or the muscles of the back may contract, causing strong arching of the vertebral column and retraction of the head.

*Tremor* may be a purely hysterical manifestation, occurring either alone or with paralysis and contracture. It most commonly involves the hands and arms; more rarely the head and legs. The movements are small and quick. In the type described by Rendu the tremor may or may not persist during repose, but it is increased or provoked by volitional movements. Volitional or intention tremor may exist, simulating closely that of insular sclerosis. Many instances of this disease are mistaken for hysteria.

(b) *Disorders of Sensation.*—Anæsthesia is most common, and usually confined to one half of the body. It may not be noticed by the patient. Usually it is accurately limited by the middle line and involves the mucous surfaces and deeper parts. The conjunctiva, however, is often spared. There may be hemianopsia. This symptom may come on slowly or follow a convulsive attack. Sometimes the various sensations are dissociated and the anæsthesia may be only to pain and to touch. The skin of the affected side is usually pale and cool, and a pin-prick may not be followed by blood. With the loss of feeling there may be a loss of muscular power. Curious trophic changes may be present, such as unilateral swelling of the hemiplegic side.

By the application of certain metals, the anæsthesia or analgesia can be transferred to the other side of the body. This may be caused by the electro-magnet and by wood and other agents, and is an effect of suggestion.

*Hyperæsthesia.*—Increased sensitiveness and pains occur in various parts. One of the most frequent complaints is of pain in the head, usually over the sagittal suture, less frequently in the occiput. This is described as agonizing, and is compared to the driving of a nail into the part; hence the name *clavus hystericus*. Neuralgias are common. Hyperæsthetic areas exist on the thorax and abdomen, pressure upon which may cause minor manifestations or even a convulsive attack. Increased sensitiveness in the ovarian region is not peculiar to hysteria. Pain in the back is an almost constant complaint with sensitive-

ness limited to certain spinous processes or diffuse. In hysterical women the pains in the abdomen may simulate those of gastric ulcer, or the condition may be almost identical with that of peritonitis; more rarely the abdominal pains resemble those of appendix disease.

*Special Senses.*—Disturbances of taste and smell are not uncommon and may cause much distress. Of ocular symptoms, retinal hyperæsthesia is common, and the patients prefer to be in a darkened room. Retraction of the field of vision is common and usually follows a convulsive seizure. It may persist for years. The color perception may be normal even with complete anæsthesia. Hysterical deafness may be complete and alternate or come on with hysterical blindness. Hysterical amaurosis may occur in children. One must distinguish between functional loss of power and simulation.

(c) *Visceral Manifestations.*—*Respiratory Apparatus.*—Of disturbances in the respiratory rhythm, the most frequent, perhaps, is an exaggeration of the deeper breath, which is taken normally every fifth or sixth inspiration, or there may be a "catching" breathing, such as is seen when cold water is poured over a person. In hysterical *dyspnœa* there is no special distress and the pulse is normal. In the syndrome of Briquet there are shortness of breath, suppression of the voice, and paralysis of the diaphragm. The anhelation is extreme. In rare instances there is bradypnœa. *Aphonia* is frequent and may persist for months or years without other special symptoms. Spasm of the muscles may occur with violent inspiratory efforts and great distress, and lead to cyanosis. Hiccough, or sounds resembling it, may be present for weeks at a time. Among the most remarkable respiratory manifestations are the hysterical cries. These may mimic the sounds produced by animals, such as barking, mewling, or grunting, and in France epidemics of them have been observed. Attacks of gaping, yawning, and sneezing may also occur.

The hysterical *cough* is a frequent symptom, particularly in young girls. It may occur in paroxysms, but is often a dry, persistent, croaking cough, extremely monotonous and unpleasant. Sir Andrew Clark called attention to a loud, barking cough occurring about the time of puberty, chiefly in boys belonging to neurotic families. The attacks, which last about a minute, recur frequently. A form of hysterical *hæmoptysis* may be deceptive and lead to a diagnosis of pulmonary disorder. The sputum is a pale-red fluid, not so bright in color as in ordinary hæmoptysis, and probably comes from the mouth or pharynx.

*Digestive System.*—Disturbed or depraved appetite, dyspepsia, and gastric pains are common. The patient may have difficulty in swallowing, apparently from spasm of the gullet. There are instances in which the food seems to be expelled before it reaches the stomach. In other cases there is incessant gagging. The *globus hystericus* is probably due to spasm of the œsophagus. In the hysterical vomiting the food is regurgitated without much effort and without nausea. This feature may persist for years without great disturbance of nutrition. The most striking and remarkable digestive disturbance in hysteria is the *anorexia nervosa* described by Sir William Gull. "To call it loss of appetite—anorexia—but feebly characterizes the symptom. It is rather an annihilation of appetite, so complete that it seems in some cases impossible ever to eat again. Out of it grows an antagonism to food which results at last and in its worst forms in spasm on the approach of food, and this in turn

gives rise to some of those remarkable cases of survival for long periods without food" (Mitchell). There are three special features in anorexia nervosa. *First*, and most important, a psychical state, usually depressant, occasionally excited and restless. It is not always hysterical. *Secondly*, stomach symptoms, loss of appetite, regurgitation, vomiting, and the whole series of phenomena associated with nervous dyspepsia. *Thirdly*, emaciation, which reaches a grade seen only in cancer and dysentery. The patient finally takes to bed, and in extreme cases lies upon one side with the thighs and legs flexed, and contractures may occur. Food is either not taken at all or only upon urgent compulsion. The skin becomes wasted, dry, and covered with bran-like scales. No food may be taken for several weeks at a time, and attempts to feed may be followed by severe spasms. Although the condition looks so alarming, these patients, when removed from their home surroundings and treated by isolation, sometimes recover in a remarkable way. It may take many months before any improvement is noted. Death, however, may follow with extreme emaciation. In one fatal case the girl weighed only 49 pounds. No lesions were found post mortem.

Hysterical *tympanites* is common, caused usually by tonic contraction of the diaphragm and retraction of the abdominal muscles. It may be associated with peristaltic unrest. Frequent movements may be due to disturbance in the small or large bowel. An obstinate form of diarrhoea is found in some hysterical patients, which proves very intractable and is associated especially with the taking of food. It seems an aggravated form of the looseness of bowels to which many nervous people are subject on emotion or of the tendency which some have to diarrhoea immediately after eating. An entirely different form is that produced by what Mitchell called the irritable rectum, in which scybala are passed frequently, sometimes with great violence. Constipation is more frequent and may be due to lack of attention to the need for defecation or to spasm (vagotonia). In extreme cases the bowels may not be moved for two or three weeks. Other disturbances are ano-spasm or intense pain in the rectum apart from any fissure. Hysterical ileus and faecal vomiting are among the most remarkable of hysterical phenomena. Following a shock there are constipation, tympanites, vomiting, sometimes hæmatemesis. The constipation grows worse, everything taken by the mouth is rejected, the vomitus becomes faecal in character, even scybala are brought up, and suppositories and enemata are vomited. The symptoms may continue for weeks and then gradually subside. Laparotomy—even thrice in one patient—has shown a perfectly normal-looking condition of the bowels (Parkes Weber).

*Cardio-vascular*.—Rapid action of the heart on slight emotion, with or without the subjective sensation of palpitation, is often a source of great distress. A slow pulse is less frequent. Pains about the heart may simulate angina. Flushes in various parts are common. Sweating may occur, or the *seborrhœa nigricans*, causing a darkening of the skin of the eyelids.

Among the more remarkable vaso-motor phenomena are the so-called stigmata or hamorrhages in the skin, such as were present in the celebrated case of Louise Lateau. In many cases these are undoubtedly fraudulent, but if, as appears credible, such bleeding may occur in the hypnotic trance, there seems no reason to doubt its possibility in the trance of religious ecstasy.

(d) *Joint Affections*.—To Sir Benjamin Brodie and Sir James Paget we

owe the recognition of these extraordinary manifestations. Perhaps no single affection has brought more discredit upon the profession, for the cases are very refractory, and often fall into the hands of a charlatan or faith-healer, under whose touch the disease may disappear at once. Usually it affects the knee or the hip, and may follow a trifling injury. The joint is usually fixed, sensitive, and swollen. The surface may be cool, but sometimes the local temperature is increased. To the touch it is very sensitive and movement causes great pain. In protracted cases the muscles are somewhat wasted, and in consequence the joint looks larger. The pains are often nocturnal, at which time the local temperature may be increased. While, as a rule, neuromimetic joints yield to proper management, there are instances in which organic change followed the functional disturbance. Intermittent hydrarthrosis may be a manifestation of hysteria, sometimes with transient paresis.

(e) *Mental Symptoms*.—Mental perversions of all kinds are common in hysterical patients and not much dependence can be placed on statements either about themselves or about others. A morbid craving for sympathy may lead to the commission of all sorts of bizarre and foolish acts.

Hallucinations and delirium may alternate with emotional outbursts of an aggravated character. There is a condition which may be spoken of as the *status hystericus*. For weeks or months they may be confined to bed, entirely oblivious to their surroundings, with a delirium sometimes associated with unpleasant objects. The nutrition may be maintained, but there is a heavy, foul breath. With seclusion and care recovery usually takes place within three or four months. At the onset of these attacks and during convalescence the patients must be incessantly watched, as a suicidal tendency is not uncommon.

Of hysterical manifestations in the higher centres that of *trance* is the most remarkable. This may develop spontaneously without any convulsive seizure, but more frequently it follows hysteroid attacks. Catalepsy may be present, a condition in which the limbs are plastic and remain in any position in which they are placed.

(f) *Manifestations*.—(1) *Edema*. Puffiness of the face, even unilateral, and swelling of the hands are not uncommon and the features of Raynaud's disease may be met with. A white and a blue type of edema is recognized, and either may be associated with paralyses, motor and sensory. (2) *Stigmata*. Local bleedings have been described, sometimes, as in the so-called marks of the cross, on forehead, hands, feet and side, as in the famous case of Louise Lateau. Organic lesions of the skin (blisters) are claimed to have been produced by hypnotic suggestion (Hadfield) and the stigmata are probably produced by auto-suggestion in the trance state. (3) *Pathomimia*, the self-inflicted injuries, usually of the skin, by caustics, etc. In a case seen at the Hôtel-Dieu with Dieulafoy, the patient, supposed to be the subject of severe trophic disorder, submitted to amputation of the arm before a confession was obtained that the lesions were self-inflicted.

(g) *Hysterical Fever*.—In hysteria the temperature, as a rule, is normal. The cases with fever may be grouped as follows: (1) Instances in which the fever is the sole manifestation. These are rare, but there are cases in which the chronic course, the retention of nutrition, and the entirely negative condition of the organs leaves no other diagnosis possible. In one case the patient had for four or five years an afternoon rise of temperature, usually to 102°

or 103°. She was well nourished and had no pronounced hysterical symptoms, beyond the interrupted sighing respiration so often seen.

(2) Cases of fever with spurious local manifestations. These are very troublesome and deceptive cases. The patient may be suddenly taken ill with pain in various regions and elevation of temperature. The case may simulate meningitis. There may be pain in the head, vomiting, contracted pupils, and retraction of the neck—which may persist for weeks—and some anomalous manifestation during convalescence may alone indicate to the physician that he has had to deal with hysteria, and has not, as he perhaps flattered himself, cured a case of meningitis. There is an hysterical pseudo-phthisis with pain in the chest, slight fever, and the expectoration of a blood-stained mucus. The cases of hysterical peritonitis may show fever.

(3) *Hyperpyrexia*.—It is a suggestive fact that the cases of paradoxical temperatures in which the thermometer has registered 112° to 120° have been in women. Fraud has been practised in nearly all these cases.

**Astasia; Abasia.**—These terms, indicating respectively inability to stand and inability to walk, have been applied by Charcot and Blocq to conditions characterized by loss of the power of standing or of walking, with retention of muscular power, co-ordination, and sensation. Blocq's definition is as follows: "A morbid state in which the impossibility of standing erect and walking normally is in contrast with the integrity of sensation, of muscular strength, and of the co-ordination of the other movements of the lower extremities." The condition forms a symptom group and is a functional neurosis. Knapp analyzed 50 cases, of which half were in women. In 21 cases hysteria was present; in 3, chorea; in 2, epilepsy; and in 4, intention psychoses. As a rule, the patients, though able to move the feet and legs perfectly when in bed, are either unable to walk properly or can not stand at all. The disturbances are very varied, and different forms have been recognized. The commonest, according to Knapp's analysis, is the paralytic, in which the legs give out as the patient attempts to walk and "bend under him as if made of cotton. There is no rigidity, no spasm, no inco-ordination. In bed, sitting, or even while suspended, the muscular strength is found to be good." Other cases are associated with spasm or ataxia; there may be movements which stiffen the legs and give to the gait a somewhat spastic character. In others there are sudden flexions of the legs, or of the arms, or a saltatory, spring-like soasm. The condition is a manifestation of hysteria.

**Diagnosis.**—Inquiry into the occurrence of previous manifestations and the mental conditions may give important information. These questions, as a rule, should not be asked the mother, who of all others is least likely to give satisfactory information. The occurrence of the globus hystericus, of emotional attacks, of weeping and crying is always suggestive. The points of difference between the convulsive attacks and true epilepsy may give difficulty at first. The hysterical paralyses are very variable and apt to be associated with anæsthesia. The contractures may be deceptive, but the occurrence of areas of anæsthesia, of retraction of the visual field, and the development of minor hysterical manifestations give valuable indications. The contractures disappear under full anæsthesia. Special care must be taken not to confound the spastic paraplegia of hysteria with lateral sclerosis.

The visceral manifestations are usually recognized without much difficulty.

The practitioner has constantly to bear in mind the strong tendency in hysterical patients to practise deception.

**Treatment.**—The prophylaxis may be gathered from the remarks on the relation of education to the disease. The successful treatment of hysteria demands qualities possessed by few physicians. The first element is a due appreciation of the nature of the disease on the part of the physician and friends. It is pitiable to think of the misery which has been inflicted on these unhappy victims by the harsh and unjust treatment which has resulted from false views of the nature of the trouble; on the other hand, worry and ill health, often the wrecking of mind, body, and estate, are entailed upon the near relatives in the nursing of a protracted case. The minor manifestations, attacks of the vapors, the crying and weeping spells, are not of much moment and rarely require treatment. The physical condition should be carefully looked into and the mode of life regulated so as to insure system and order in everything. A congenial occupation offers the best remedy for many of the manifestations. Any functional disturbance should be attended to and tonics prescribed. Special attention should be paid to the bowels.

**PSYCHOTHERAPY**, in which the important features are hypnosis, analysis, suggestion, and reëducation.

**Hypnosis.**—The majority of hysterical patients can be hypnotized, but the general opinion of those who know most on the subject is that by hypnosis alone hysteria is rarely cured. Sometimes a brilliant miracle is wrought but as a routine treatment it has fallen into disfavor even in France.

**Suggestion.**—Babinski defines suggestion as "the action by which one endeavors to make another accept or realize an idea which is manifestly unreasonable." On the other hand, persuasion is applied when the ideas are reasonable, or at least are not in opposition to good sense. Most writers, however, use the word "suggestion" as meaning the introduction of mental associations and modifications of the patient's mental state leading to betterment. In proper hands it is a most powerful instrument, particularly when the patient has faith in the person who makes it. After a careful and sympathetic examination and testing the electrical reactions of the muscles of a paralyzed limb the suggestion to the hysteric, "Now you will be able to move it" may be all-sufficient. A strong, imperative command may have the same effect.

**Re-education.**—In both hysteria and neurasthenia this should be the aim of all reasonable practice, but it is not always feasible: some of our patients would have to be rebuilt from the blastoderm. With patience and method much may be done, and the special merit of Weir Mitchell's work and of his system (which is not simply a rest cure, as many suppose) is that it is an elaborate plan of re-education. The essentials are that the patient should be isolated from her friends and under the charge of an intelligent nurse. The physical condition is carefully studied and a rigid daily régime carried out: A milk diet of three to four quarts daily, rising to five or six, varying the food as the patient improves, and as the weight increases. This may be followed by a rapid gain in weight and the disappearance of the unpleasant abdominal symptoms. Massage, hydrotherapy, and electricity are adjuncts, but very much depends upon the tact, patience, and, above all, the personality of the physician; *the man counts more than the method.* The mental condition has

to be carefully studied and the patient's attitude toward life influenced by specially selected literature, careful conversation, and suggestion.

**THE ANALYTICAL METHOD.**—Introduced by Breuer and extended by Freud, it is partly the method of the confessional, in which the sinner poured out his soul in the sympathetic ear of the priest, but it also enables the patient to bring out into the open what he may not consciously know. It is a difficult procedure, not for all to attempt, exhausting alike to patient and doctor, and, when thoroughly carried out, time consuming. In the hands of those who have practised it skilfully, very good results have been obtained, particularly in young and carefully selected cases. This statement of the method is taken from Jelliffe (*System of Medicine*, 2nd Ed., Vol. V.) :

"His (Freud's) general procedure is to place the patient in a recumbent position, the physician sitting behind the patient's head at the end of the lounge. The physician thus remains practically out of sight of the patient, who is then asked to give a detailed account of his troubles, and to say everything that comes to the mind irrespective of its seeming logic or sense, and apart from disturbing, mortifying, or unwise suggestions. In all such histories gaps are inevitable. These the patient is urged to fill in by thinking closely of the attendant circumstances, speaking aloud all of the fitting thoughts that pass during this search ('free association'). All the thoughts are requested to be uttered, notwithstanding their disagreeable nature. The patient must exercise no critique and remain passive. It will be found that the disagreeable thoughts are pushed back with the greatest resistance. This is made all the more striking since the hysterical reaction, i. e., the symptom, is the symbolic expression of the realization of a repressed wish and gives the patient some gratification. A great effort is made to retain the symptom, especially as its origin is not really perceived, and since it represents, in symbol, the individual's former conscious strivings. In psycho-analysis one attempts to overcome all of these resistances, and by a series of judicious and tactful probings reconduct into the patient's consciousness the hidden thoughts which underlie these symptoms. Every symptom has some meaning; behind it there lies some associated mechanism, the origin of which the patient unconsciously or partly consciously represses. In the psycho-neurotic symbol may be read the cryptic expression of the original thought driven back and hidden.

"To slowly analyze and pick apart the mechanism is the object of the analytical method. One needs not only special tact for such excursions into the subtleties of the mental life of some individuals, but also a developed method of interpretation. Every act, every symbolic expression or action, lapse in speech, mannerism, needs to be carefully noted and its bearing co-ordinated. Freud lays particular emphasis on the analysis of dreams, since he believes that in the dream the subconscious, or the 'repressed conscious' is more apt to reveal itself.

"It is of the utmost importance to trace back into the earliest years the striking emotional influences that have come into experience, as, for Freud, the hysterical reaction consists in a perverted type of reaction to these experiences. As is known, the blurring, or loss of an emotional influence—an affect, in short—is due to a number of factors. In normal life forgetting is the commonest type of a corrective adaptation, and forgetting is carried out with special ease if the emotional stress has not been excessive. Forgetting,

however, is only a secondary phenomenon, and usually is more successful if the immediate reaction has been an adequate one. Such immediate reactions express themselves as tears, as anger, as impulsive acts, etc., and in such reactions the affect is discharged. In every-day life one calls it giving vent to one's feelings. If, however, the reaction is suppressed, the effect becomes united to the memory of the experience, and an emotional complex, or, to use a rather broad simile, a psychic boil, results, which must heal by absorption, by discharge, or by other means. Freud uses the term *ab-react* (*abreagieren*) to signify the adequate reaction, or discharge, of such effects or their resulting complexes. Talking the whole thing over, giving vent to one's secrets and confessions are well-known *abreactions*.

"In hysteria certain of these complexes remain prominent; they are neither reacted to promptly, nor is their unpleasant feeling tone diminished by the blurring process of forgetting, although it is characteristic of the Freud point of view that the actual experience which gives rise to them becomes forgotten and the cause of the affect disturbance which becomes later converted, it may be into physical signs, remains apparently unknown to the patient. It must be dug out by psycho-analysis, and when once discovered catharsis takes place and the patient becomes cured."

HYPROTHERAPY is of great value, especially wet packs, salt baths, and various douches. General tonics, such as arsenic and iron, may be helpful, especially if the patients are nervous and anæmic. Sedatives are rarely indicated. Occasionally bromides may be necessary, but for the relief of sleeplessness all possible measures should be resorted to before the employment of drugs. The wet pack given hot or cold at night will usually suffice.

## X. NEURASTHENIA

(*Psychasthenia*)

**Definition.**—A condition of weakness or exhaustion of the nervous system, giving rise to various forms of mental and bodily inefficiency.

The term, an old one, popularized by Beard, covers an ill-defined, motley group of symptoms, which may be either general and the expression of derangement of the entire system, or local and limited to certain organs.

**Etiology.**—The causes may be grouped as hereditary and acquired.

(a) **HEREDITARY.**—We do not all start in life with the same amount of nerve capital. Parents who have led irrational lives, indulging in excesses of various kinds, or who have been the subjects of nervous complaints or of mental trouble, may transmit to their children an organization which is defective in what, for want of a better term, we must call "nerve force." Such individuals start handicapped with a neuropathic predisposition, and furnish a considerable proportion of our neurasthenic patients. As Van Gieson sonorously puts it, "the potential energies of the higher constellations of their association centres have been squandered by their ancestors." So long as these individuals are content to transact a moderate business with their life capital, all may go well, but there is no reserve, and in the exigencies of modern life these small capitalists go under and come to us as bankrupts.

(b) **ACQUIRED.**—The functions, though perverted most readily in persons who have inherited a feeble organization, may also be damaged in persons with no neuropathic predisposition by use which is excessive in proportion to the strength—i. e., by strain. The cares and anxieties attendant upon the gaining of a livelihood may be borne without distress, but in many persons the strain becomes excessive and is first manifested as *worry*. The individual loses the distinction between essentials and non-essentials, trifles cause annoyance, and the entire organism reacts with unnecessary readiness to slight stimuli, and is in a state which the older writers called “irritable weakness.” If such a condition be taken early and the patient given rest, the balance is quickly restored. In this group may be placed a large proportion of the neurasthenia which we see in professional and business men. Neurasthenia may follow the infectious diseases, particularly influenza, typhoid fever, and syphilis. The abuse of alcohol or tobacco may predispose to neurasthenia.

(c) **SEXUAL CAUSES.**—Undoubtedly the part played in the production of neurosis by sex factors is of the first importance. As stated, Freud regards sexual trauma as the basis of hysteria, and he also regards neurasthenia as largely a product of disturbance in the sexual sphere. For him and his school the sex impulses furnish the basis of the psychoneuroses. Repressed as they have to be in so many in our modern civilization, without normal outlet, the thought formations, retained in the unconscious state, express themselves by means of somatic phenomena—the objective features of hysteria and neurasthenia. *Cherchez la femme* is a safe rule in investigating a neurotic male. Freud may have ridden his hobby too hard, particularly in the insistence upon the importance of infantile sexuality, but in recognizing the rôle of the younger Aphrodite in the lives of men and women he has but followed the great master, Plato, who saw, while he deplored, the havoc wrought by her universal dominance.

**Symptoms.**—These are extremely varied, and may be general or localized; more often a combination of both. The appearance of the patient is suggestive, sometimes characteristic, but difficult to describe. Important information can be gained by the physician if he observes the patient closely as he enters the room—the way he is clothed, his manner, his facial expression, and the humor which he is in. Loss of weight and slight anæmia may be present. The debility may reach a high grade and the patient be confined to bed. Mentally the patients are usually low-spirited and despondent; women are frequently emotional. Fatigue is an important feature.

The *local* symptoms may dominate the situation, and there have accordingly been described a whole series of forms of the disease—cerebral, spinal, cardio-vascular, gastric, and sexual. In all there is a striking lack of accordance between the symptoms of which the patient complains and the objective changes discoverable by the physician. In nearly every clinical form the predominant symptoms are referable to pathological sensations and the psychic effects of these. Imperfect sleep is complained of by a majority of patients, or, if not complained of, is found to exist on inquiry.

In the *cerebral* or psychic form the symptoms are chiefly connected with an inability to perform the ordinary mental work. Thus, a row of figures can not be correctly added, the dictation or the writing of a few letters is a source of the greatest worry, the transaction of petty details is a painful effort, and

there is loss of power of fixed attention. With this condition there may be no headache, the appetite may be good, and the patient may sleep well. As a rule, however, there are sensations of fulness and weight or flushes, if not actual headache. Insomnia is frequent and may be the first manifestation. Some patients are good-tempered and cheerful but a majority are moody, irritable, and depressed.

*Hyperæsthesia*, especially to sensations of pain, is one of the main characteristics of almost all neurasthenic individuals. The sensations are nearly always referred to some special region—the skin, eyes, joints, blood-vessels, or viscera. It is frequently possible to localize a number of points painful to pressure (Valleix's points). In some patients there is marked vertigo, occasionally resembling that of Ménière's disease.

If such pathological sensations continue for a long time the mood and character of the patient gradually alter. The so-called "irritable humor" develops. Many obnoxiously egoistic individuals met with in daily life are in reality examples of psychic neurasthenia. Complaint is made of everything. The patient demands the greatest consideration for his condition; he feels that he has been insulted if his desires are not always immediately granted, but at the same time he has but little consideration for others. Indeed, in the severer forms he may show a malicious pleasure in attempting to make people who seem happier than himself uncomfortable. Such patients complain frequently that they are "misunderstood" by their fellows.

In many cases the so-called "anxiety conditions" gradually come on; one scarcely ever sees a case of advanced neurasthenia without some form of anxiety or fear. In the simpler forms there may be a fear of impending insanity, of approaching death or of apoplexy. More frequently the anxious feeling is localized somewhere in the body—in the præcordial region, the head, the abdomen, the thorax, or more rarely in the extremities. In some cases the anxiety becomes intense and the patients are restless, and declare that they do not know what to do with themselves. They may throw themselves upon a bed, crying and complaining, and making convulsive movements with the hands and feet. Suicidal tendencies are not uncommon in such cases, and the patients may in desperation take their own lives.

Involuntary mental activity may be very troublesome; the patient complains that when he is overtired thoughts which he can not stop or control run through his head with lightning-like rapidity. In other cases there is marked absence of ideas, the individual's mind being so filled up owing to the overexcitability of latent memory pictures that he is unable to form the proper associations for ideas called up by external stimuli. Sometimes a patient complains that a definite word, a name, a number, or a song keeps running in his head in spite of all he can do to abolish it.

In the severer cases the so-called "*phobias*" are common. A frequent form is *agoraphobia*, in which patients when they come into an open space are oppressed by a feeling of anxiety. They seem "frightened to death," and commence to tremble; they complain of compression of the thorax and palpitation of the heart. They may break into profuse perspiration and assert that they feel as though chained to the ground or that they can not move a step. It is remarkable that in some such cases the open space can be crossed if the individual be accompanied by some one, even by a child, or if he carry

a stick or an umbrella! Other people are afraid to be left alone (monophobia), especially in a closed compartment (claustrophobia).

The fear of people and of society is known as anthropophobia. A whole series of other phobias has been described—batophobia, or the fear that high things will fall; pathophobia, or fear of disease; siderodromophobia, or fear of a railway journey; siderophobia or astrophobia, fear of thunder and lightning. Occasionally we meet with individuals who are afraid of everything and every one—victims of the so-called pantophobia. By psycho-analysis it is possible to explain the mechanism of these fears.

The *special senses* may be disturbed, particularly vision. An aching or weariness of the eyeballs after reading a few minutes or flashes of light are common symptoms. The "irritable eye," the so-called nervous or neurasthenic asthenopia, is familiar to every physician. There may be acoustic disturbances—hyperalgnesia and even true hyperacusia.

A common symptom is *pressure in the head* which, variously described, may be diffuse, but is more frequently referred to some one part, especially the occipital region. When *spinal symptoms* predominate, in addition to many of the features mentioned, the patients complain of weariness on the least exertion, of weakness, pain in the back, intercostal neuralgiform pains, and of aching pains in the legs. There may be spots of local tenderness on the spine. The pain may be spontaneous, or noticed only on pressure or movement. Occasionally there may be disturbances of sensation, particularly numbness and tingling, and the reflexes may be increased. Visceral neuralgias, especially in connection with the genital organs, are frequent. An aching pain in the back or in the back of the neck is the most constant complaint. In women it is often impossible to say whether the condition is neurasthenia or hysteria. It is in these cases that the disturbances of muscular activity are most pronounced, and in the French writings *amyosthenia* plays an important rôle.

The symptoms may be irritative or paretic, or a combination of both. Disturbances of coördination are not uncommon in the severer forms. These are particularly prone to involve the associated movements of the eye muscles, leading to asthenopic lack of accommodation. Drooping of one eyelid is common, probably owing to insufficient innervation on the part of the sympathetic rather than to paresis of the oculomotor nerve. Occasionally Romberg's symptom is present, and the patient, or his physician, may fear a beginning tabes. More rarely there is disturbance of such co-ordinated acts as writing and articulation, not unlike those at the onset of general paresis. Such symptoms are always alarming, and the greatest care must be taken in establishing a diagnosis. That they may be the symptoms of pure neurasthenia can not be doubted.

The *reflexes* are usually increased, the deep reflexes especially never being absent. The conditions of the superficial reflexes is less constant, though these, too, are usually increased. The pupils are often dilated, and the reflexes are usually normal. There may be inequality of the pupils. Errors in refraction are common, the correction of which may give great relief.

In another form the muscular weakness is extreme and very thorough examination is necessary before deciding as to the nature of the affection, since in some instances serious mistakes have been made. Here belong the *atremia*

of Nettel, the *akinesia algera* of Möbius, and the neurasthenic form of *actasia abasia* described by Binswanger.

In other cases the *cardio-vascular* symptoms are the most distressing, and may occur with only slight disturbance of the cerebro-spinal functions, though the conditions are nearly always combined. Palpitation of the heart, irregular and very rapid action (neurasthenic tachycardia), and pains and oppressive feelings in the cardiac region are the most common. Some of these are due to the "dropped" heart which may be dilated. The slightest excitement may be followed by increased action of the heart, sometimes associated with sensations of dizziness and anxiety, and the patients fear that they suffer from serious disease of this organ. Attacks of "pseudo-angina" may occur.

*Vaso-motor disturbances* are marked in many cases. Flushes of heat, especially in the head, and transient hyperæmia of the skin may be very distressing symptoms. Profuse sweating may occur, local or general, and sometimes nocturnal. The pulse may show interesting features, owing to the extreme relaxation of the peripheral arterioles. The arterial throbbing may be almost as marked as in aortic insufficiency. The pulse may have a somewhat collapsing quality and the capillary pulse may be seen. A characteristic symptom in some cases is the *throbbing aorta*. This "preternatural pulsation in the epigastrium," as Allan Burns calls it, may be extremely forcible and suggest aneurism. The associated subjective sensations may be very unpleasant, particularly when the stomach is empty.

In women especially, and sometimes in men, the peripheral blood-vessels are contracted, the extremities are cold, the nose is red or blue, and the face has a pinched expression. These patients feel much more comfortable when the cutaneous vessels are distended, and resort to various means to favor this (wearing of heavy clothing, use of diffusible stimulants).

The general features of *gastro-intestinal neurasthenia* have been dealt with under the section on nervous dyspepsia. The connection with dilatation of the stomach, floating kidney, and *enteroptosis* has been mentioned.

In *sexual neurasthenia* there is an irritable weakness of the sexual organs manifested by nocturnal emissions, depression after intercourse, and often by a dread of impotence. The mental condition of these patients is pitiable, and they fall an easy prey to quacks and charlatans. In males these symptoms are frequently due to diseased conditions in the deep urethra, especially of the verumontanum, and prostate. Spermatorrhœa is the bugbear of the majority. They complain of losses especially with defecation or micturition. Microscopic examination sometimes reveals the presence of spermatozoa. Actual nervous impotence is not uncommon. The "painful testicle" is a well-known neurasthenic phenomenon. In severer cases, especially those bearing the stigmata of degeneration, there may be sexual perversion. In females it is common to find a tender ovary, and painful or irregular menstruation. There may be disturbances in the sex sphere.

**Diagnosis.**—*Psychasthenia*.—Under this term Janet would separate from neurasthenia the cases characterized by mental, emotional, and psychical disturbances, imperative ideas, phobias of all sorts, doubts, enfeebled will, uncontrollable movements, and many of the borderland features of the insanity of young persons. It is really an inherited psychoneurosis, while neurasthenia is usually acquired. Obsessions of all sorts characterize the condition and

there may be a feeling of unreality and even of loss of personality. How complicated the condition may be is shown from the following varieties distinguished by Janet: (1) The *doubter*, in whom obsessive ideas are not very precise, more of the nature of a general indication rather than a specific idea, such as a craze for research, for explanation, for computing. (2) The *scrupulous*, whose obsessions are of a moral nature. Their manias are of literalness of statement, of exact truth, of conjuration, of reparation, of symbols, etc. (3) The *criminal*, whose obsessive ideas are of homicide, theft, and other overt acts. The impulsive idea is stronger in this than in the other varieties. (4) The *inebriates*, morphinomaniac, etc., in whom the impulse seems to be least resistible. (5) The *genesically perverted*. (6) *Delirious psychasthenia*, in which a delirious state of mind occurs, connected with the obsession.

Neurasthenia, above all other diseases, has to be diagnosed from the subjective statements of the patient, and from a study of his general behavior rather than from the physical examination, which, however, is of the highest importance in excluding other diseases likely to be confounded with it. That somatic changes occur and that physical signs are often to be made out is very true, but there is nothing typical or pathognomonic in these objective changes.

The *hypochondriac* differs from the neurasthenic in the excessive psychic distortion of the pathological sensations to which he is subject. He is the victim of actual delusions regarding his condition.

The confusion of neurasthenia with *hysteria* is still more frequent; in women especially a diagnosis of hysteria is often made when in reality the condition is neurasthenia. In the absence of hysterical paroxysms, of crises, and of those marked emotional and intellectual characteristics of the hysterical individual the diagnosis of hysteria should not be made. If hysterical stigmata (paralyses, convulsions, contractures, anæsthesias, alterations in the visual field, etc.) are present, the diagnosis is not difficult.

*Epilepsy* is not likely to be confounded with neurasthenia if there be definite epileptic attacks, but the cases of *petit mal* may be puzzling.

The onset of *exophthalmic goitre* may be mistaken for neurasthenia, especially if there be no exophthalmos at the beginning. The emotional disturbances and the irritability of the heart may mislead the physician. *Tuberculosis* should always be excluded and careful search made for signs of any *internal secretion* disturbance. In pronounced cases of neurasthenia the differential diagnosis from the various psychoses may be extremely difficult.

The two forms of *organic disease* of the nervous system with which neurasthenia is most likely to be confounded are tabes and general paresis. The symptoms of the spinal form of neurasthenia may resemble those of tabes, while the symptoms of the psychic form of neurasthenia may be very similar to those of general paresis. The diagnosis, as a rule, presents no difficulty if the physician makes a thorough routine examination. It is only a superficial study that is likely to lead one astray. In tabes a consideration of the sensory disturbances, of the deep reflexes, and of the pupillary findings will establish the presence or absence of the disease. In general paresis there is sometimes more difficulty. The onset is often characterized by symptoms quite like those of neurasthenia, and the physician may overlook the grave nature of the malady. The mistake in the other direction is, however, perhaps just as common. A physician who has seen a case of general paresis arise out of what

appeared to be pronounced neurasthenia is too prone afterward to suspect every neurasthenic to be developing the malign affection. The most marked symptoms of psychic exhaustion do not justify a diagnosis of general paresis even when the history is suspicious, unless along with it there is a definite paresis of the pupils, of the facial muscles, or of the muscles of articulation. The physician should be sharply on the lookout for intellectual defects, paraphasia, facial paresis, and sluggishness of the pupils. The examination of the spinal fluid will remove any doubt.

**Treatment.**—**PROPHYLAXIS.**—Many patients come under our care a generation too late for satisfactory treatment, and it may be impossible to restore the exhausted capital. The greatest care should be taken in the rearing of children of neuropathic predisposition. From a very early age they should be submitted to a process of "psychic hardening," every effort being made to strengthen the bodily and mental condition. Even in infancy the child should not be pampered. Later on the greatest care should be exercised with regard to food, sleep, and school work. Complaints of children should not be too seriously considered. Much depends upon the example set by the parents. An emotional, constantly complaining mother will rack the nervous system of a delicate child. In some instances, for the welfare of a developing boy or girl, the physician may find it necessary to advise removal from home.

Neurotic children are especially liable during development to fits of temper and of emotional disturbance. These should not be too lightly considered. Above all, violent chastisement in such cases is to be avoided, and loss of temper on the part of the parent or teacher is particularly pernicious for the nervous system of the child. Where possible, in such instances, the best treatment is to put the obstreperous child immediately to bed, and if the excitement and temper continue a warm bath followed by a cool douche may be effective. If he be put to bed after the bath sleep soon follows.

Special attention is necessary at puberty in both boys and girls. If there be at this period any marked tendency to emotional disturbance or to intellectual weakness the child should be removed from school and every care taken to avoid unfavorable influences.

**PERSONAL HYGIENE.**—Throughout life individuals of neuropathic predisposition should obey scrupulously certain hygienic and prophylactic rules. Intellectual work especially should be judiciously limited and alternate frequently with periods of repose. Excitement of all kinds should be avoided, and such individuals do well to be abstemious in the use of tobacco, tea, coffee, and alcohol, if, indeed, they be permitted to use them at all. The habit of taking at least once a year a prolonged holiday should be urgently enjoined upon every neuropathic individual. In many instances it gives great relief and rest if the patient can take his holiday away from his relatives.

During ordinary life nervous people should, during some portion of each day, pay rational attention to the body. Cold baths, swimming, exercises in the gymnasium, gardening, golf, lawn tennis, cricket, hunting, shooting, rowing, and bicycling are of value in maintaining the general health. Such exercises are to be recommended only to individuals physically equal to them. If neurasthenia is well established the greatest care must be observed in the ordering of exercise. Many nervous girls have been completely broken down by following injudicious advice with regard to exercise.

**TREATMENT.**—The treatment of neurasthenia when once established presents a varied problem to the thoughtful physician. Every case must be handled upon its own merits, no two, as a rule, requiring exactly the same methods. In general it will be the aim to remove the patient as far as possible from the influences which have led to his downfall, and to restore to normal the nervous mechanisms which have been weakened by injurious influences. The general character of the individual, his physical and social status, must be considered and the therapeutic measures carefully adjusted to these.

The diagnosis having been settled, the physician may assure the patient that with prolonged treatment, during which his coöperation is absolutely essential, he may expect to get well. He must be told that much depends upon himself and that he must make a vigorous effort to overcome certain of his tendencies, and that all his strength of will will be needed to further the progress of the cure. In business or professional men, in whom the condition develops as a result of overwork or overstudy, it may be sufficient to enjoin absolute rest with change of scene and diet. A trip abroad or, if there are symptoms of nervous dyspepsia, a residence at one of the spas will usually prove sufficient. The excitement of large cities should be avoided. The longer the disease has lasted and the more intense the symptoms have been, the longer the time necessary for the restoration of health. In cases of any severity the patient must be told that at least six months' complete absence from business, under strict medical guidance, will be necessary. Shorter periods may be of benefit, which, however, as a rule, will be only temporary.

It will often be found advisable to make out a daily programme, which shall occupy almost the whole time of the patient. At first he need know nothing about this, the care being given over entirely to the nurse. As improvement advances, moderate physical and intellectual exercises, alternating frequently with rest and the administration of food, may be undertaken. Some one hour of the day may be left free for reading, correspondence, conversation, and games. In some instances the writing of letters is particularly harmful and must be prohibited or limited. Cultured individuals may find benefit from attention to drawing, painting, modelling, translating, the making of abstracts, etc., for short periods in the day.

In some cases, including a large proportion of neurasthenic women, a systematic rest treatment rigidly carried out should be tried. The patient must be isolated and all regulations must be strictly adhered to, the consent of the patient and the family having first been gained. The treatment of the gastric and intestinal symptoms has been considered. For the irregular pains, particularly in the back and neck, the cautery is invaluable.

*Hydrotherapy* is indicated in nearly every case if it can be properly applied. Much can be done at home or in a hospital, but for systematic hydrotherapeutic treatment residence in a suitable sanitarium is better. The wet pack is of especial value and, particularly at night, is perhaps the best remedy against insomnia we have. Salt baths are more helpful to some patients. The various forms of douches, partial packs, etc., may be valuable in individual cases. Electrotherapy is of some value, though only in combination with psychic treatment and hydrotherapy.

Special care should be given to the recognition of local disease and proper measures instituted. Attention to the eyes is important. Infection of the

naso-pharynx, teeth or tonsils, sinus disease, visceroptosis, or anæmia should be corrected. In women the pelvic organs, and in men the deep urethra and prostate may require treatment.

Treatment by drugs should be avoided as much as possible. They are of benefit chiefly in the combating of single symptoms. Alcohol, morphia, opium, or cocaine should never be given. General tonics may be helpful, especially if the individual be anæmic, when arsenic and iron are indicated. For the severer pains and nervous attacks some sedative may be necessary, especially at the beginning of the treatment. The bromides may be given with advantage. An occasional dose of phenacetine or acetyl-salicylic acid may be required, but the less of these substances we use the better. For the relief of sleeplessness all possible measures should be resorted to before the employment of drugs. The wet pack will usually suffice. If necessary to give a drug, sulphonal, trional, barbital, or other hypnotics may be employed.

PSYCHOTHERAPY.—Hypnotism is rarely indicated. Carefully practised suggestion is most helpful and psycho-analysis is of value.

The use of religious ideas and practices has come into vogue in various forms, as Christian Science, Mental Healing, etc. It is an old story. In all ages, and in all lands, the prayer of faith, to use the words of St. James, has healed the sick; and we must remember that amid the Æsculapian cult, the most elaborate and beautiful system of faith healing the world has seen, scientific medicine took its rise. As a profession, consciously or unconsciously, more often the latter, *faith* has been one of our most valuable assets and Galen expressed a great truth when he said, "He cures most successfully in whom the people have the greatest confidence." It is in these cases of neurasthenia and psychasthenia, the weak brothers and the weaker sisters, that the personal character of the physician comes into play, and once let him gain the confidence of the patient, he can work the same miracles as Our Lady of Lourdes or Ste. Anne de Beaupré. Three elements are necessary: first, a strong personality in whom the individual has faith—Christ, Buddha, Æsculapius (in the days of Greece), one of the saints, or, what has served the turn of common humanity very well, a physician. Secondly, certainly accessories—a shrine, a sanctuary, the services of a temple, or for us a hospital or its equivalent, with a skillful nurse. Thirdly, suggestion, either of the "only believe," "feel it," "will it" attitude of mind, which is the essence of every cult and creed, or of the active belief in the assurance of the physician that health is within reach.

## XI. THE TRAUMATIC NEUROSES

(*Railway Brain and Railway Spine; Traumatic Hysteria*)

**Definition.**—A morbid condition following shock which presents the symptoms of neurasthenia or hysteria or of both.

Erichsen regarded the condition as the result of inflammation of the meninges and cord, and gave it the name "railway spine." Walton and J. J. Putnam, of Boston, were the first to recognize the hysterical nature of many of the cases, and to Westphal's pupils we owe the name traumatic neurosis.

**Etiology.**—The condition follows an accident, often in a railway train, in which injury has been sustained, or succeeds a shock or concussion, from which the patient may apparently not have suffered in his body. A man may appear well for several days, or even a week or more, and then the symptoms appear. Bodily shock or concussion is not necessary. The affection may follow a profound mental impression; thus, an engine-driver ran over a child, and received thereby a very severe shock, subsequent to which the most pronounced symptoms of neurasthenia developed. Severe mental strain with bodily exposure may cause it, as in a naval officer who was wrecked in a violent storm and exposed for more than a day in the rigging before he was rescued. A slight blow, a fall from a carriage or on the stairs may suffice. The possibility of actual injury of the spine should always be considered.

**Symptoms.**—The cases may be divided into three groups: simple neurasthenia, cases with marked hysterical manifestations, and cases with severe symptoms indicating or simulating organic disease.

(1) **SIMPLE TRAUMATIC NEURASTHENIA.**—The first symptoms usually develop a few weeks after the accident, which may or may not have been associated with an actual trauma. The patient complains of headache and tired feelings. He is sleepless and finds himself unable to concentrate his attention upon his work. A condition of nervous irritability develops, which may have a host of trivial manifestations, and the entire mental attitude of the person may be changed. He dwells constantly upon his condition, becomes despondent and low-spirited, and in extreme cases melancholia may develop. He may complain of numbness and tingling in the extremities, and in some cases of much pain in the back. The bodily functions may be well performed, though such patients usually have, for a time at least, disturbed digestion and loss in weight. The physical examination may be entirely negative. The reflexes are slightly increased, as in ordinary neurasthenia. The pupils may be unequal; cardio-vascular changes may be present in a marked degree.

(2) **CASES WITH MARKED HYSTERICAL FEATURES.**—Following an injury of any sort, neurasthenic symptoms, like those described above, may develop, and in addition symptoms regarded as characteristic of hysteria. The emotional element is prominent, and there is but slight control over the feelings. The patients have headache, backache, and vertigo. A violent tremor may be present, and constitute the most striking feature. In an engineer who developed subsequent to an accident a series of nervous phenomena the most marked feature was an excessive tremor of the entire body, which was specially manifest during emotional excitement. The most pronounced hysterical symptoms are the sensory disturbances. As first noted by Putnam and Walton, hemianæsthesia may occur as a consequence of trauma. This is a common symptom in France, but rare in England and the United States. Achromatopsia may exist on the anæsthetic side. A second, more common, manifestation is limitation of the field of vision, similar to that in hysteria.

(3) **CASES IN WHICH THE SYMPTOMS SUGGEST ORGANIC DISEASE.**—As a result of spinal concussion, without fracture or external injury, symptoms may subsequently develop suggestive of organic disease, which may come on rapidly or at a late date. In a case reported by Leyden the symptoms following the concussion were at first slight and the patient was regarded as a simulator, but finally the condition became aggravated and death resulted. The post-

mortem showed a chronic pachymeningitis, doubtless from the accident. The cases in this group about which there is so much discussion are those which display marked sensory and motor changes. Following an accident in which the patient has not received external injury a condition of excitement may develop within a week or ten days; he complains of headache and backache, and sensory disturbances are found, either hemianæsthesia or areas in which the sensation is much benumbed; or painful and tactile impressions may be distinctly felt in certain regions, and the temperature sense is absent. The distribution may be bilateral and symmetrical in limited regions or hemiplegic in type. Limitation of the field of vision is usually marked, and there may be disturbance of the senses of taste and smell. The superficial reflexes may be diminished; usually the deep reflexes are exaggerated. The pupils may be unequal; the motor disturbances are variable. The French writers describe cases of monoplegia with or without contracture, symptoms upon which Charcot laid great stress as a manifestation of profound hysteria. The combination of sensory disturbances with paralysis, particularly if monoplegic, and the occurrence of contractures without atrophy and with normal electrical reactions, may be regarded as distinctive of hysteria.

In rare cases following trauma and succeeding to symptoms which may have been regarded as neurasthenic or hysterical there are organic changes which may prove fatal. That this occurs is demonstrated clearly by post mortem examinations. The features upon which the greatest reliance can be placed as indicating organic change are optic atrophy, bladder symptoms, particularly in combination with tremor, paresis, and exaggerated reflexes.

The anatomical changes are not very definite. When death follows spinal concussion within a few days there may be no apparent lesion, but in some instances the brain or cord showed punctiform hæmorrhages. Edes reported 4 cases in which degeneration in the pyramidal tracts followed concussion or injury of the spine; but in all these cases there was marked tremor and the spinal symptoms developed early, or followed immediately upon the accident.

**Diagnosis.**—A condition of fright and excitement following an accident may persist for days or even weeks, and then gradually pass away. The symptoms of neurasthenia or of hysteria which subsequently develop present nothing peculiar and are identical with those which occur under other circumstances. Care must be taken to recognize simulation, and, as in these cases the condition is largely subjective, this is sometimes extremely difficult. In a careful examination a simulator will often reveal himself by exaggeration of certain symptoms, particularly sensitiveness of the spine, and by increasing voluntarily the reflexes. Mankopff suggests as a good test to take the pulse rate before, during, and after pressure upon an area said to be painful. If the rate is quickened, it is held to be proof that the pain is real. This is not, however, always the case. It may require careful study to determine whether the individual is honestly suffering from the symptoms of which he complains. A still more important question is, Has the patient organic disease? The symptoms given under the first two groups of cases may exist in a marked degree and may persist for several years without the slightest evidence of organic change. Hemianæsthesia, limitation of the field of vision, monoplegia with contracture, may all be present as hysterical manifestations, from which recovery may be complete. The diagnosis of an organic lesion should be limited to those cases

in which optic atrophy, bladder-troubles, and signs of sclerosis of the cord are well marked—indications either of degeneration of the lateral columns or of multiple sclerosis. Examination by the X-rays is an important aid and has showed in some cases definite injury to the spine.

**Prognosis.**—A majority of patients with traumatic hysteria recover. In railway cases, so long as litigation is pending and the patient is in the hands of lawyers, the symptoms usually persist. Settlement is often the starting-point of a speedy and perfect recovery. On the other hand, there are a few cases in which the symptoms persist even after the litigation has been closed; the patient goes from bad to worse and psychoses develop, such as melancholia, dementia, or occasionally progressive paresis. And, lastly, in extremely rare cases organic lesions may result as a sequence.

The function of the physician acting as medical expert in these cases consists in determining (*a*) the existence of actual disease, and (*b*) its character, whether simple neurasthenia, severe hysteria, or an organic lesion. The outlook for ultimate recovery is good except in cases which present the more serious symptoms above mentioned. Nevertheless traumatic hysteria is one of the most intractable affections which we are called upon to treat. In the treatment of the traumatic neuroses the practitioner may be guided by the principles laid down for the treatment of hysteria and neurasthenia.

## L. VASO-MOTOR AND TROPHIC DISEASES

### I. RAYNAUD'S DISEASE

**Definition.**—A vascular change, without organic disease of the vessels, chiefly seen in the extremities, but occurring also in the internal parts, in which a persistent ischæmia or a passive hyperæmia leads to disturbance of function or to loss of vitality with necrosis.

**Etiology.**—It is a comparatively rare disease. There were only 19 cases in about 20,000 medical patients admitted to the Johns Hopkins Hospital. Women are more frequently attacked than men—62.5 to 37.5 per cent. in Monro's series. Sixty per cent. of the cases occurred in the second and third decades, but no age is exempt. A case has been reported in a six-months-old child and in a woman of 77 years.

Several members of a family may be affected. Neurotic and hysterical patients are more prone to the disease. Damp and cold weather appears to favor its occurrence. Severe chilblain leading to superficial necrosis represents a type of the malady. In the infectious diseases areas of gangrene occur, but such cases should not be included under Raynaud's disease, nor should the local gangrene associated with arteritis. Syphilis is suggested in some cases.

**Pathology.**—According to the definition, cases are excluded in which organic disease of the vessels is present. In advanced cases sclerosis of the blood-vessels has been found; and neuritis has been described, but neither is an essential factor. Changes in the spinal cord have been reported, but in a majority of cases the examination has been negative. The local ischæmia is

an expression of a constrictor influence causing spasm of the arteries and arterioles, so that not a drop of blood enters a part. This may be followed in an hour or two, or less, by active *hyperæmia*; the arteries and arterioles dilate widely and the dead-white finger becomes a bright pink. While *hyperæmia* may follow the *ischæmia* directly, more commonly there is an intervening period of *asphyxia* in which the finger becomes blue. In frost-bite, active *hyperæmia*, cyanosis, and *ischæmia* is the order. In Raynaud's disease the order is usually *ischæmia*, *asphyxia*, and *hyperæmia*. In frost-bite it seems clear that the *asphyxia* is due to a backward flow from the veins, to which the *ischæmia* yields as the part thaws, before the arteries passing to the part can be felt to pulsate. The *asphyxia* of Raynaud's disease may be due to the same cause; contraction of the veins has been seen by Barlow and by Weiss, but that was when *asphyxia* already existed. The first thing must be the relaxation of the spasm of the venules and veins to permit the blood to enter the empty capillaries. In moderate grades of *asphyxia* some little blood trickles through the sluice gates, but in the deep purple skin of a typical example of Raynaud's disease the circulation has ceased and death of the part is imminent. The necrosis is a simple matter, as simple as if a string is tied tightly about the finger.

The disease is the result of some as yet unknown instability of the vaso-motor system.

**Symptoms.**—There are various grades of the disease, of which mild, moderate, and severe types may be recognized. In the *mild* forms the disease never gets beyond the stage of such vascular disturbance as is frequently seen in chilblains. The hands alone may be affected. In the winter, on the slightest exposure, there is acro-cyanosis, which gives place in the warmth to active *hyperæmia*, sometimes with swelling, throbbing, and aching. The so-called "beefsteak" hand is often a great annoyance to women. It is a vaso-motor disturbance representing a potential case of Raynaud's disease. In these mild attacks one finger may be white and the adjacent ones red and blue.

The condition may persist for years and never pass on to necrosis. In a case of *moderate* severity a woman, aged say twenty or twenty-five, after a period of worry or ill health, has pains in the fingers, or a numbness or tingling; then she notices that they are white and cold, and in an hour or so they become red and hot. Within a day or two a change occurs; they remain permanently blue perhaps as far as the second joint or to the knuckles. There is pain, sometimes severe enough to require morphia. The cyanosis persists and the tip of one finger or the terminal joint of another gets darker and a few blebs form. The other fingers show signs of restored circulation, but necrosis has occurred in the pad of one finger and perhaps the terminal inch of another. The necrotic parts gradually separate, and the patient may never have another attack, or in a year or two there is a recurrence.

The *severe* form is a terrible malady, and may affect fingers and toes at once and sometimes the tip of the nose and the ears. The pain is of great severity. Both feet may be swollen to the ankle with the toes black. It may look as if both feet would become gangrenous, but as a rule the process subsides, and in a case even of great severity only the tips of the toes are lost. A severe attack may last for months, when the patient recovers with

the loss of two or three fingers or toes, a snip off the edge of both ears and a scar on the tip of the nose. Such attacks may occur year by year, and there are terrible instances in which the patients have lost both hands and feet.

Of the parts affected Monro states that in 43 per cent. of the cases one or both of the upper extremities is involved. Parts other than the extremities may be attacked, as the chin, lips, nates, and eyelids.

**Complications.**—Temporary amblyopia due to spasm of the retinal vessels, transient aphasia, and transient hemiplegia have been met with. In one case there were three attacks of aphasia with hemiplegia with complete recovery. Associated with these were the features of Raynaud's disease. The patient died in a severe attack with pain in the right hand, gangrene to the elbow, and coma. Epilepsy occurs in a number of cases, and in one case (Thomas) the attacks only occurred in the winter when he had Raynaud's disease.

*Albuminuria* may occur during the attacks. *Hæmoglobinuria* occurs in some cases, and was studied by the well-known surgeon, Druitt, in his own case. It is of the same nature as the paroxysmal hæmoglobinuria.

*Scleroderma* of the fingers may follow recurring attacks. Occasionally true generalized scleroderma begins with the features of Raynaud's disease. Arthritis has been present in certain cases.

**Diagnosis.**—There is rarely any difficulty in the diagnosis. One condition closely simulates it, namely, local gangrene of the toes associated with obliterative arteritis; but this occurs most frequently in older persons, in diabetic subjects, or in connection with well-marked arterio-sclerosis. As a rule, the pulse in such cases is not to be felt in the dorsal artery. Allied to this form is an affection described by Buerger, thrombo-angiitis obliterans. In the early stages the resemblance to Raynaud's disease is very close. In acute infections, particularly typhus fever, occasionally in typhoid fever and malaria, areas of multiple gangrene occur. The distribution is usually different, and there is rarely any difficulty in distinguishing this form.

There are cases of multiple neurotic skin gangrene in hysterical and nervous patients, in the majority of which the lesions are self-inflicted. In military recruits local gangrene of the big toe has been caused by carbolic acid, and many of these lesions are simulated.

**Treatment.**—In many cases the attacks recur for years uninfluenced by treatment. In the forms of local asphyxia in the feet, the patient should be kept in bed with the legs elevated. The toes should be wrapped in cotton wool. The pain is often intense and may require morphia. Carefully applied, systematic massage of the extremities is sometimes beneficial. Galvanism may be tried. Nitroglycerin has been warmly recommended. Calcium lactate in 15 grain (1 gm.) doses, three or four times a day, is sometimes very effectual. It often relieves chilblains. Small doses of thyroid extract or anterior lobe pituitary gland substance (gr. ii, 0.12 gm.) sometimes are useful. Cushing introduced a plan of treatment with the tourniquet which has proved successful in several cases. An elastic bandage, or, better, a pneumatic tourniquet, is applied to the extremity tight enough to shut off the arterial circulation and left for some minutes. On releasing the constriction the member flushes brightly, owing to the vaso-motor relaxation. The application in cases of severe spasm may have to be repeated at frequent

intervals before the vascular constriction in the affected parts is overcome, and the normal temperature and color return.

## II. ERYTHROMELALGIA

(*Red Neuralgia*)

**Definition.**—"A chronic disease in which a part or parts—usually one or more extremities—suffer with pain, flushing, and local fever, made far worse if the parts hang down" (Weir Mitchell). The name signifies a painful, red extremity.

Mitchell speaks of it as a "painful nerve-end neuritis." Dehio suggests that there may be irritation in the cells of the ventral horns of the cord at certain levels. Excision of the nerves passing to the parts has been followed by relief. In one of Mitchell's cases gangrene of the foot followed excision of four inches of the musculo-cutaneous nerve and stretching of the posterior tibial. Sclerosis of the arteries was found. Of 9 cases in which the local conditions were studied anatomically, the only constant change was a chronic endarteritis (Beatty Shaw).

**Symptoms.**—In 1872 (*Phila. Med. Times*, November 23d), in a lecture on certain painful affections of the feet, Weir Mitchell described the case of a sailor, aged forty, who after an African fever began to have "dull, heavy pains, at first in the left and soon after in the right foot. There was no swelling at first. When at rest he was comfortable and the feet were not painful. After walking the feet were swollen. They scarcely pitted on pressure, but were purple with congestion; the veins were everywhere singularly enlarged, and the arteries were throbbing visibly. The whole foot was said to be aching and burning, but above the ankle there was neither swelling, pain, nor flushing." As the weather grew cool he got relief but no treatment seemed to benefit him. This brief summary of Mitchell's first case gives an accurate picture of the disease. His second communication, "On a Rare Vaso-Motor Neurosis of the Extremities," appeared in the *American Journal of the Medical Sciences* for July, 1878, while in his *Clinical Lessons on Nervous Diseases*, 1897, will be found additional observations.

The disease is rare. The feet are much more often affected than the hands. The pain may be of the most atrocious character. It is usually, but not always, relieved by cool weather; in one case winter aggravated the trouble. Cold applications, faradism and massage have been employed in treatment. Excision of nerves and amputation have been done.

## III. ANGIO-NEUROTIC OEDEMA

(*Quincke's Disease*)

**Definition.**—An affection characterized by the occurrence of local oedematous swellings, more or less limited in extent, and of transient duration. Severe colic is sometimes associated.

**Etiology.**—There is a marked hereditary predisposition, sometimes through many generations, transmitted by males and females. In one family in five generations 22 members were affected (Osler). The sexes are affected equally and the disease occurs most often in the second, third and fourth decades. Infections, nervous influences, trauma and cold are assigned as causes. It often occurs with urticaria.

**Symptoms.**—The oedema appears suddenly and is usually circumscribed. It may appear in the face; the eyelid is a common situation; or it may involve the lips or cheek. The backs of the hands, the legs, or the throat may be attacked. Usually it is transient, associated perhaps with slight gastro-intestinal distress, and the affection is of little moment. There may be a remarkable periodicity in the outbreak of the oedema. In Matas' case this was very striking; the attack came on every day at eleven or twelve o'clock. The swellings appear in various parts; only rarely are they constant in one locality. The hands, face, pharynx, glottis, and genitalia are most frequently affected. Itching, heat, redness, or in some instances urticaria, may precede the outbreak. Sudden oedema of the larynx may prove fatal. Two members of one family died of this complication. The lower lip may be swollen to such a degree that the mouth can not be opened. The hands enlarge suddenly, so that the fingers can not be bent. The attacks recur every three or four weeks. Accompanying them are usually gastro-intestinal attacks, severe colic, pain, nausea, and sometimes vomiting. It is possible that some of the cases of Leyden's intermittent vomiting belong to this group. The colic is of great intensity and usually requires morphia. Arthritis apparently does not occur. Periodic attacks of cardialgia may also occur with the oedema. Hæmoglobinuria has occurred in several cases. There is a hysterical variety in which the oedema affects geometrical areas, has abrupt edges and may be accompanied by sensory disturbances but not by gastro-intestinal attacks.

The disease has affinities with urticaria, the giant form of which is probably the same disease, and with Henoch's purpura. Quincké regards the condition as a vaso-motor neurosis, with which the permeability of the vessels is suddenly increased. The prognosis is good in the non-hereditary forms; the duration varies from one to three years. A large proportion of deaths occurs from oedema of the larynx.

The **treatment** is unsatisfactory. In cases with anæmia and general nervousness, tonics, particularly large doses of strychnia, do good. Improvement may follow the prolonged use of nitroglycerin; calcium lactate may be tried, in doses of 15 grains (1 gm.) thrice daily. Epinephrine, 1-1000 solution, (m vii, 0.5 c. c.) given hypodermically and repeated in fifteen minutes has been helpful. In cases due to food susceptibility, a small dose of the causal material may be given an hour before the usual feeding is taken. In one case in which there was susceptibility to any albumin, the administration of peptone was successful.

#### IV. ERYTHROEDEMA

(*Acrodynia: "Pink Disease"*)

To this disease other names have been given, e.g., epidemic erythema, erythroedema, polyneuritis, and dermato-polyneuritis. It is difficult to know where to place it. Swift, of Adelaide, Australia, drew attention to it in 1914. Since then cases have been reported from many countries, with some difference in the main features. The disease occurs at ages from four months to four years. There is a change in disposition with irritability and discomfort; the child is fretful and miserable. Photophobia, anorexia, weakness, slight fever occasionally, and increased pulse rate occur. The *rash* is important and is variable in character, sometimes like "prickly heat." It may fade and reappear. Erythema and eczema-like are used by some to describe it. The exact color varies from pink to a dark red. There are marked itching and excessive perspiration. The hands and feet are red, swollen and cold, are compared to raw beef, and show desquamation. Stomatitis and gingivitis with loss of the teeth may be present. There may be loss of hair and the nails may fall off. Disturbance of sensation is common and there is a varying degree of hypotonia and pseudo-paresis. Peripheral neuritis and changes in the spinal cord and nerve roots have been found. The children are subject to respiratory disorders. A leucocytosis is common. The mortality is about 5 or 6 per cent. The *diagnosis* should be made from the peculiar features. The *treatment* is symptomatic.

#### V. PERSISTENT HEREDITARY OEDEMA OF THE LEGS

(*Milroy's Disease*)

This remarkable condition, first described by Milroy of Omaha, is characterized by persistent oedema of the legs, without any traceable cause or any constitutional features. It affects males and females equally. In Milroy's series 22 persons were affected among 97 in six generations; in Hope and French's series 13 of 42 persons in five generations. The oedema is strictly limited to the lower limbs and varies very slightly. In some instances there are remarkable acute attacks, with chill, fever, and increase of swelling. Except mechanically the condition does not seriously interfere with health. Constant support by bandages is advisable.

There is a remarkable familial affection described by Edgeworth of Bristol (*Lancet*, July 22, 1911), with a general subcutaneous oedema. Of six infants born of healthy parents, all but one died within the first few months, with general oedema, following upon diarrhoea. The cases differ essentially from those of oedema neonatorum.

## VI. FACIAL HEMIATROPHY

A rare affection characterized by progressive wasting of the bones and soft tissues of one side of the face. The atrophy starts in childhood, but in a few cases has not come on until adult life. Perhaps after a trifling injury or disease the process begins, either diffusely or more commonly at one spot on the skin. It gradually spreads, involving the fat, then the bones; more particularly the upper jaw and last and least the muscles. The wasting is sharply limited at the middle line, and the appearance of the patient is very remarkable, the face looking as if made up of two halves from different persons. There is usually change in the color of the skin and the hair falls. Owing to the wasting of the alveolar processes the teeth become loose and drop out. The eye on the affected side is sunken, owing to loss of orbital fat. There is usually hemiatrophy of the tongue on the same side. Disturbance of sensation and muscle twitching may precede or accompany the atrophy. In a majority of cases the atrophy has been confined to one side of the face, but there are instances in which the disease was bilateral, and a few cases in which there were areas of atrophy on the back and arm of the same side.

In Mendel's case the terminal stage of an interstitial neuritis was found in all the branches of the trigeminus, from its origin to the periphery, most marked in the superior maxillary branch.

The disease is recognized at a glance. The facial asymmetry associated with congenital wryneck must not be confounded with progressive facial hemiatrophy. Other conditions to be distinguished are: Facial atrophy in poliomyelitis, and in the hemiplegia of infants and adults; the atrophy following nuclear lesions and sympathetic nerve paralysis; acquired facial hemi-hypertrophy, which may by contrast give to the other side an atrophic appearance; and scleroderma, if confined to one side of the face. The precise nature of the disease is doubtful, but it is a suggestive fact that in many cases the atrophy followed the acute infections. It is incurable.

## VII. SCLERODERMA

**Definition.**—A condition of localized or diffused induration of the skin.

**Varieties.**—Two forms are recognized; the circumscribed, which corresponds to the keloid of Addison and to morphea, and the diffuse, in which large areas are involved.

**Etiology.**—The disease affects females more frequently than males and occurs most commonly at the middle period of life. *Scleroderma neonatorum* is a different affection. The disease is more common in the United States than statistics indicate. The senior author saw 20 cases in sixteen years. It is regarded as a tropho-neurosis, as due to changes in the arteries of the skin, and to endocrine disturbance, especially of the thyroid, adrenal and pituitary glands. The thyroid has been found atrophied.

**Pathology.**—There is pigment deposit in the epidermis with an increased amount of connective tissue in the corium and subcutaneous tissues. The

superficial vessels are contracted and the deeper vessels are surrounded by leucocytes.

**Symptoms.**—In the *circumscribed form* there are patches, from a few centimetres in diameter to the size of the hand or larger, in which the skin has a waxy or dead-white appearance, and to the touch is brawny, hard, and inelastic. Sometimes there is a preliminary hyperæmia and subsequently there are changes in color, either areas of pigmentation or of complete atrophy of the pigment—leucoderma. The sensory changes are rarely marked. The secretion of sweat is diminished or entirely abolished. The areas are situated most frequently about the breasts and neck, sometimes in the course of the nerves. The patches may develop with great rapidity and persist for months or years; sometimes they disappear in a few weeks.

The *diffuse form*, though less common, is more serious. It begins in the extremities or in the face, and the patient notices that the skin is unusually hard and firm, or that there is a sense of stiffness or tension in making movements. Gradually the skin becomes firm and hard, and so united to the subcutaneous tissues that it cannot be picked up or pinched. It may look natural, but more commonly is glossy, drier than normal, and unusually smooth. With reference to the localization, in Lewin and Heller's statistics in 66 cases the disease was universal; in 203, regions of the trunk were affected; in 193, parts of the head or face; in 287, portions of one or other of the upper extremities; and in 122, portions of the lower extremities. In 80 cases there were disturbances of sensation. The disease may gradually extend and involve the skin of an entire limb. When universal, the face is expressionless, the lips can not be moved, mastication is hindered, and it may become extremely difficult to feed the patient. The hands become fixed and the fingers immobile, on account of the extreme induration of the skin over the joints. Remarkable vaso-motor disturbances are common, as extreme cyanosis of the hands and legs. Tachycardia may be present. The disease is chronic, lasting for months or years and there are instances of its persistence for more than twenty years. Recovery may occur or the disease may be arrested. One patient, with extensive involvement of the face, ears, and hands, improved very much. The patients are apt to succumb to pulmonary complaints or nephritis. Arthritis has been noted in some instances; in others, endocarditis. Raynaud's disease may be associated with it. The *pigmentation* of the skin may be as deep as in Addison's disease, for which cases have been mistaken; *scleroderma* may occur with exophthalmic goitre.

The remarkable atrophy known as *sclerodactylia* belongs to this disorder. There are *symmetrical* involvements of the fingers, which become deformed, shortened, and atrophied; the skin becomes thickened, of a waxy color, and is sometimes pigmented. Multiple subcutaneous nodules, not unlike tophi, but not uratic, occur about the fingers. Bullæ and ulcerations have occurred and great deformity of the nails. The disease has usually followed exposure; the patients are much worse during the winter, and are curiously sensitive to cold. There may be changes in the skin of the feet, but deformity similar to that in the hand has not been noted. Some patients present in addition diffuse sclerodermatous changes of the skin of other parts. In Lewin and Heller's monograph there are 35 cases of isolated sclerodactylism, and 106 cases in which it was combined with scleroderma.

**Treatment.**—The patients require to be warmly clad and to be guarded against exposure, as they are particularly sensitive to cold. Warm baths followed by frictions with oil should be systematically used. Thyroid feeding should be tried thoroughly in the diffuse form. In one case the disease appeared to be arrested; the patient took the extract for seven years. In a second case, after a year the face became softer, and there was permanent improvement. In a case of extensive localized scleroderma the patches became softer and the pigmentation much less intense. Salol in 15 grain (1 gm.) doses three times a day is stated to have been helpful. The views as to the value of X-ray treatment are conflicting.

### VIII. AINHUM

This is a disease of the fifth, rarely of the fourth and other toes in which a groove forms at the digito-plantar fold and deepens until the toe drops off. Described first by Da Silva Lima in 1852, it has since been met with in many tropical regions, in the southern states of America, and very rarely in temperate regions, as Canada and Italy. Nothing has been determined as to the *etiology*. It may occur in families, and is more common in males. There is endarteritis, with proliferation of the epidermis. Parasites have not been found. It is a local disease without symptoms and in the affected toe there is no disability and rarely any pain, except when the skin of the groove ulcerates. The toe drops off in about two years. It is said that the affection may occur in the fingers. A longitudinal section across the groove will sometimes stop the progress, otherwise the toe should be removed.

### IX. LIPODYSTROPHIA PROGRESSIVA

A rare affection, possibly confined to females, in which the subcutaneous fat gradually disappears from the face, arms, and trunk. The cause of the condition is unknown. Beginning usually about the tenth to thirteenth year, the wasting is progressive, but limited to the parts mentioned. The buttocks and legs remain normal and by contrast look abnormally plump. The breasts are spared. In the early stages it resembles the "bilateral atrophy of the face."

## SECTION XIII

# DISEASES OF THE LOCOMOTOR SYSTEM

## A. DISEASES OF THE MUSCLES

### I. MYOSITIS

**Definition.**—Inflammation of the voluntary muscles.

A primary myositis occurs as an acute, subacute, or chronic affection. It is seen in two chief forms—the suppurative and non-suppurative.

**I. Suppurative myositis** (infectious myositis) is especially frequent in Japan, where Miyake saw 33 cases in 21 months, and took cultures from 32; in 2 cases the results were negative, but in 27 a pure culture of *Staphylococcus aureus* was obtained, while in another the streptococcus and in 2 more the albus with the aureus was grown. The malady may involve one or many muscles, and is usually sudden in its onset with high fever and marked prostration. Subsequently abscesses occur in the indurated muscles, and pyæmia may ensue if the implicated areas are not thoroughly evacuated.

**II. Dermato-myositis.**—An acute or subacute inflammation of the muscles of unknown origin associated with œdema and dermatitis. Steiner collected 28 cases from the literature and reported two cases from the Hopkins clinic. The nature of the disease is unknown. The muscle inflammation is multiple, and associated with œdema and dermatitis. The muscles are stiff and firm, but fragile, with serous infiltration, great proliferation of the interstitial tissue, and fatty degeneration. The onset is usually gradual, the muscles of the extremities being first involved and later those of the trunk. There is moderate fever with sweating and enlargement of the spleen. The skin over the affected muscles is œdematous and the dermatitis takes various forms, erythematous, urticarial or erysipelatoid. Pain is present, worse on movement or pressure, and sensory changes may occur. The course is usually progressive and after some months death may result from involvement of the muscles of deglutition and respiration, perhaps with broncho-pneumonia. The duration is usually one to three months. Of Steiner's 28 cases, 17 died. If recovery results there is atrophy of the muscles. The features suggest trichiniasis which may be distinguished by examination of a portion of muscle. The treatment is symptomatic.

**III. Polymyositis hæmorrhagica.**—This form resembles the dermatomyositis in general features, but differs in the presence of hæmorrhages into and between the muscles. Of the ten cases analyzed by Thayer four recovered. Purpura and hæmorrhages from the mucous membranes may occur.

**IV. Secondary Myositis.**—This may occur in some infections, as septicæmia and typhoid fever, and is constant in trichiniasis. In scurvy a chronic myositis may occur, sometimes associated with hæmorrhages. In chronic

alcoholism, myositis may occur with neuritis. In some cases of fibrositis there is an associated myositis, difficult to distinguish. Syphilis may cause myositis.

## II. MYOSITIS OSSIFICANS PROGRESSIVA

This is a progressive inflammatory affection of the locomotor system of unknown origin, characterized by the gradual formation of bony masses in the fasciæ, muscles, aponeuroses, tendons, ligaments, and bones, with resulting ankylosis of most of the articulations (Steiner). The process begins in the neck or back, usually with swelling of the affected muscles, redness of the skin, and slight fever, or with small nodules in the muscles which appear and disappear. After subsiding an induration remains, which becomes progressively harder as the transformation into bone takes place. The disease may ultimately involve a majority of the skeletal muscles. Microdactylism of the thumbs and big toes is present in 75 per cent. of the cases.

*Local* myositis ossificans may occur after repeated injuries or a single severe one. There are also non-traumatic and neurotic forms. It may follow an abdominal incision. The diagnosis has to be made from hæmatoma, new growth and exostoses. The history and X-ray study are important. This form usually reaches its maximum size rapidly, to remain stationary or diminish, usually the latter.

## III. FIBROSITIS

**Definition.**—An inflammation of fibrous tissue which may occur in many parts of the body, involving ligaments, tendons, muscle sheaths, fasciæ, aponeuroses, periosteum, nerve sheaths, or any part where fibrous tissue is found. It is associated with arthritis in many cases and to designate the involvement of special parts certain terms are employed, as synovitis, teno-synovitis, bursitis, and peri-neuritis. Fibrositis is very common and is responsible for many painful conditions often termed myalgia, "muscular rheumatism," neuritis, etc.

**Etiology.**—The essential cause is infection in the great majority of cases and usually from a local focus. Trauma, muscular strain and exposure to cold and wet are determining factors, especially in acute attacks. Prolonged strain on tendons or ligaments may be responsible for a chronic fibrositis, but infection is often added. It may follow exposure to a draught of air, as from an open window in a carriage or sudden chilling after heavy exertion may bring on an attack. Gouty persons seem prone to this affection. One attack renders an individual more liable to another. It is usually acute at first, but may become subacute or chronic, the last being more common in later life.

**Pathology.**—The changes are usually in the white fibrous tissue and are of an inflammatory nature. In acute cases there is a serous exudation in the affected parts and following this there may be proliferation of the fibrous tissue. This may extend between the muscle fibres and cause stiffness and pain. Disability with muscular atrophy may result from this. Nodules sometimes form which may be painful.

**Symptoms.**—In the acute forms the affection is usually local. The constitutional disturbance is slight, and, even in severe cases, there may be no fever. Pain is a prominent feature and may be constant or occur only when the structures are in certain positions or moved. It may be a dull ache, like the pain of a bruise, or sharp, severe, and cramp-like. It is often sufficiently intense to cause the patient to cry out. Pressure on the affected part usually gives relief. As a rule, the pain lasts from a few hours to a few days, although occasionally it is prolonged for weeks. It is very apt to recur.

There is a form occurring chiefly in the muscles of the head and neck, causing at first swelling and puffiness, later indurations. They are found particularly in the muscles at the back of the neck, but they are occasionally present in the muscles of the abdomen and limbs. The affection of the muscles of the head and neck may be associated with headache, the so-called *indurative headache*. Some are very similar to migraine. In the abdominal muscles these limited swellings may cause pain and suggest appendicitis.

The following are the principal varieties:

(1) "*Lumbago*," a term which means nothing more than pain in the lower back, is due to many causes. (a) *Fibrositis*, in the muscles or fibrous tissue about the spine, is a common cause and may recur at short intervals. It comes on suddenly and may incapacitate the patient, any movement, particularly stooping or turning, causing severe pain. (b) *Ischæmic lumbago*, described as a form of intermittent claudication, may be bilateral or unilateral and is excited by movement. The pain is between the twelfth rib and the crest of the ilium and may radiate forward. The area is not tender and the pain is dependent on muscular exertion. (c) *Static* conditions, due to faulty posture, which may be lateral (one leg shorter) or antero-posterior, flat feet, stooping, occupation, etc. (d) *Anatomical variations* of the transverse processes of the fifth lumbar vertebra. (e) *Arthritis of the spine*. (f) *Sacro-iliac* joint strain or relaxation. (g) *Neuritis* of the posterior nerve roots. (h) Pain due to *pelvic disease* in males (prostate, etc.) or females. (i) *Trauma*, especially with lifting in a stooped position. The diagnosis of "backache" as being due to fibrositis should only be made after other possibilities are excluded. In every case the effort should be made to arrive at an etiological diagnosis as only then is proper treatment possible. For the cases due to spondylitis or sacro-iliac joint disease some form of fixation is useful; faulty posture should be corrected and flat feet receive attention.

(2) Stiff neck or *torticollis* affects the muscles of the antero-lateral or back region of the neck. It is very common, often unilateral, and occurs most frequently in the young. The patient holds the head in a peculiar manner turned to one side, and rotates the whole body in attempting to turn it.

(3) *Pleurodynia* involves the intercostal muscles on one side, and in some instances the pectorals and serratus magnus. This is, perhaps, the most painful form of the disease, as the chest can not be at rest. A deep breath or coughing causes pain, sometimes over a very limited area. It may be difficult to distinguish from intercostal neuralgia, in which, the pain is usually more circumscribed and paroxysmal, and there are tender points along the course of the nerves. It is sometimes mistaken for pleurisy, but careful examination readily distinguishes between the affections.

(4) Among other forms are cephalodynia, affecting the muscles of the

head; scapulodynia, omodynia, and dorsodynia, affecting the muscles about the shoulder and upper part of the back. Fibrositis also occurs in the abdominal muscles, in the muscles of the extremities and about the joints. In the legs it causes tenderness and pain, increased by use. Areas of thickening or infiltration may be palpated. Nodules are sometimes felt in the sole of the foot and in the hands. The chronic forms have soreness or pain associated with varying degrees of disability. There may be marked stiffness of the muscles, which are sometimes painful on pressure and may show definite tender areas of induration. Periosteal involvement gives pain and tenderness on pressure.

**Diagnosis.**—If a careful examination is made there should be little difficulty in recognizing the surface lesions but deeper ones give difficulty. Disease of the nerve roots or viscera should be excluded.

**Treatment.**—As a focus of infection is often responsible careful search should be made for any possible infection and proper treatment given. If done early the chronic conditions are not likely to result. Rest of the affected parts is of the first importance, and it is well to protect them from cold by a covering of flannel. Fixation by strapping should be done wherever possible. No belief is more widespread among the public than in the efficacy of porous plasters for pains of all sorts. If the pain is severe and agonizing, a hypodermic of morphia gives relief. For lumbago acupuncture is, in acute cases, an efficient treatment. Needles of from three to four inches in length (ordinary bonnet-needles, sterilized, will do) are thrust into the lumbar muscles at the seat of pain, and withdrawn after five or ten minutes. In many cases the cautery gives great relief and in obstinate cases blisters may be tried. Heat or counter-irritation in any form is useful and at the outset a Turkish bath may cut short an acute attack. The bowels should be freely opened and large amounts of water taken. The salicylates are usually effectual; sodium salicylate (gr. x to xv, 0.6 to 1 gm.), acetyl-salicylic acid (gr. x, 0.6 gm.), or salol (gr. v, 0.3 gm.) may be given. Some patients respond well to colchicum (℥ xv, 1 c. c. of the wine). In chronic cases potassium iodide may be used. Persons subject to this affection should be warmly clothed, and avoid, if possible, exposure to cold and damp. Massage usually is helpful; it should be given gently at first and more vigorously later. Use of the affected parts is advisable after the acute features are over. For the lumbar form, fixation is most useful by strapping, or some form of support.

#### IV. MYOTONIA CONGENITA

(*Thomsen's Disease*)

**Definition.**—An affection characterized by tonic cramp of the muscles on attempting voluntary movements. The disease received its name from the physician who first described it, in whose family it existed for five generations.

While the disease is in a majority of cases hereditary, there are other forms of spasm very similar which may be acquired, and others still which are quite transitory. The nature of the affection is unknown. Déjerine and Sottas found hypertrophy of the primitive fibres with multiplication of the

nuclei of all the muscles, including the diaphragm, but not the heart. The spinal cord and the nerves were intact. From Jacoby's studies it is doubtful whether these changes in the muscles are characteristic or peculiar to the disease. J. Koch found, in addition to the muscle hypertrophy, degenerative and regenerative changes, which he considers sufficient to account for the myotonic disorder.

**Etiology.**—All the typical cases have occurred in family groups; a few isolated instances have been described in which similar features were present. Males are much more frequently affected; in 102 recorded cases, 91 were males and only 11 females (Hans Koch). The disease is rare in America and England; it seems more common in Germany and Scandinavia.

**Symptoms.**—The onset is in childhood. It is noticed that on account of the stiffness the children are not able to take part in ordinary games. The peculiarity is noticed only during voluntary movements. The contraction which the patient wills is slowly accomplished; the relaxation which the patient wills is also slow. The contraction often persists for a little time after he has dropped an object which he has picked up. In walking, the start is difficult; one leg is put forward slowly, it halts from stiffness for a second or two, and then after a few steps the legs become limber and he walks without any difficulty. The muscles of the arms and legs are those usually implicated; rarely the facial, ocular, or laryngeal muscles. Emotion and cold aggravate the condition. In some instances there is mental weakness. The sensation and reflexes are normal. G. M. Hammond reported three cases in one family, in which the disease began at the eighth year and was confined to the arms. It was accompanied with slight mental feebleness. The condition of the muscles is interesting. The patients appear and are muscular, and there is sometimes definite muscular hypertrophy. The force is scarcely proportionate to the size. Erb described a characteristic reaction to the electrical currents—the so-called myotonic reaction, the chief feature of which is that the contractions caused by either current attain their maximum slowly and relax slowly, and vermicular, wave-like contractions pass from the cathode to the anode. The disease is incurable, but may be arrested temporarily. No treatment is known.

## V. MYOTONIA ATROPHICA

### *(Dystrophia Myotonica)*

This is a slowly progressive form of muscular atrophy affecting special muscles, associated with delayed relaxation of muscular contractions. It was formerly regarded as an atypical form of Thomsen's disease. The disease is hereditary and familial in many cases but not in all. Males are affected more often and the onset is usually between the ages of 20 and 30. The muscles show an overgrowth of connective tissue with degeneration of the muscle fibres. No changes in the endocrine glands have been found.

**Symptoms.**—The onset is insidious, usually with the myotonia preceding the atrophy but the reverse may occur. The myotonia is usually most evident in the hands. The atrophy appears in one of several groups of muscles:

(1) The head and neck group, especially the muscles of the eyelids and the sterno-mastoids, (2) the muscles of the forearms, and (3) the anterior and lateral muscles of the legs, especially the quadriceps extensor and vasti. The inability to close the eyes, the hanging lower lip and the curious facial expression, if myotonia is present, give a characteristic appearance. There are many associated conditions. *Cataract* develops in one-third of the cases, usually matures rapidly and may occur in otherwise healthy members of the family. Loss of hair, changes in the teeth and skin, emaciation, loss of tendon reflexes, and sensory disturbances may follow. The *diagnosis* from Thomsen's disease is made by the muscular atrophy and by the dystrophic features. The *course* is prolonged and in some the progress stops. The treatment is directed to the general health and to the relief of symptoms.

## VI. PARAMYOCLONUS MULTIPLEX

(*Essential Myoclonia*)

**Definition.**—An affection described by Friedreich, characterized by irregular clonic contractions, chiefly of the muscles of the extremities, occurring either constantly or in paroxysms.

**Etiology.**—The subjects are usually degenerate young males. Hysteria, emotion, fright, trauma, sexual excesses and parathyroid disease have been suggested as possible causes. The disease may occur in several generations.

**Pathology.**—The nature of the disease is unknown. Pierce Clark suggests that it is an abiotrophy of the *corpus striatum*—the region of control of the automatic and associated movements.

**Symptoms.**—The characteristic features are the short, sudden and lightning-like contractions, not rhythmic and of equal intensity, and without the synergic quality of a purposive movement. The two sides may be unequally involved, and single muscles may be affected. The face and fingers are not often affected. Sensation is not involved; the reflexes are increased. The movements are usually absent during sleep.

**Diagnosis.**—The disease may be confounded with the various symptomatic myoclonias seen in the tics, chronic chorea, and the rhythmic movements associated with mid-brain lesions. The myoclonia with epilepsy (Unverricht) is described elsewhere.

**Treatment.**—Various forms of suggestion may be tried, but the disease may prove very resistant. The movements may cease spontaneously.

## VII. MYASTHENIA GRAVIS

(*Asthenic Bulbar Paralysis; Erb-Goldflam's Symptom-Complex*)

**Definition.**—A disease with fatigue symptoms referable to the muscular system, due to failure of innervation without definite changes in muscles or nerves.

Of 180 cases collected by McCarthy, 83 were males and 96 females. In

women the disease usually occurs before the age of twenty-five, in males in middle life. Of 56 autopsies since 1901, in 17 there was hyperplasia or persistence of the thymus and in 10 a thymic tumor, one of which was malignant. Examination of the nervous system has revealed no abnormality. Hun, Bloomer, and Streeter described an infiltration of the muscles and of the thymus gland with lymphoid cells and a proliferation of the glandular elements of the thymus. Disturbance in the lymphatic system has been found. The excretion of creatinin is decreased.

**Symptoms.**—The striking feature is weakness of the voluntary muscles, rapidly produced by exertion and disappearing with rest. Ptosis is the first manifestation in many cases. The muscles innervated by the bulb are first affected—those of the eyes, face, of mastication, and of the neck. After effort the muscles show fatigue, and if persisted in they fail to act and paresis or complete paralysis follows. All the voluntary muscles may become involved. After rest the power is recovered. In severe cases paralysis may persist. The *myasthenic reaction* of Jolly is the rapid exhaustion of the muscles, by faradism, not by galvanism. There are marked remissions and fluctuations in the severity of the symptoms. Probably there are many unrecognized mild cases.

The *diagnosis* is made from the ptosis, the facial expression, the nasal speech, the rapid fatigue of the muscles, the myasthenic reaction, the absence of atrophy, tremors, etc., and the remarkable variations in the intensity of the symptoms. Of 180 collected cases 72 proved fatal; sudden death may occur, sometimes from paralysis of the respiratory muscles. The patient may live many years and recovery take place. In *treatment*, absolute rest, both general and local, is important. Muscular fatigue should be avoided. Strychnine should be given in large doses hypodermically. Alternate courses of iodide of potassium and mercury may be tried.

## VIII. AMYOTONIA CONGENITA

(*Oppenheim's Disease*)

A congenital affection characterized by general or local hypotonus of the voluntary muscles. Oppenheim gave the name *myatonia*, but this is so similar to *myotonia* (Thomsen's disease) that the name *amyotonia* of English writers is preferable. Collier and Wilson give the following definition: "A condition of extreme flaccidity of the muscles, associated with an entire loss of the deep reflexes, most marked at the time of birth and always showing a tendency to slow and progressive amelioration. There is great weakness, but no absolute paralysis of any of the muscles. The limbs are most affected; the face is almost always exempt. The muscles are small and soft, but there is no local wasting. Contractures are prone to occur in the course of time. The faradic excitability in the muscles is lowered and strong faradic stimuli are borne without complaint. No other symptoms indicative of lesions of the nervous system occur."

Faber collected 115 cases (1917). Recovery has not been reported, but improvement was noted in 41 cases. The post-mortem changes are variable

—increase in the muscle nuclei, and in the fat and connective tissue; defective myelinization of the peripheral nerves; absence of the nerve endings in the muscle fibres, without changes in the central nervous system. The disease seems to come in the group of Gowers' abiotrophies—a failure in the proper development of the lower motor neurone.

## B. DISEASES OF THE JOINTS

### I. ARTHRITIS DEFORMANS

**Definition.**—A disease of the joints, the result of infection, characterized by changes in the synovial membranes, cartilage, and peri-articular structures, and in some cases by atrophic and hypertrophic changes in the bones. A tendency to a chronic course is the rule.

Long believed to be associated with gout and rheumatism, this relationship is disproved. There is a difference of opinion as to whether there are two distinct diseases or varying forms of the same disease included under this heading. Those who hold the former view consider that in one disease the synovial membranes and the peri-articular tissues are particularly affected (rheumatoid arthritis) and in the other disease the cartilage and bone (osteo-arthritis). The disease is common and to it belong many of the cases termed "chronic rheumatism."

**Etiology.**—**Age.**—A majority of the cases are between the ages of twenty and fifty. In A. E. Garrod's analysis of 500 cases there were only 25 under twenty years of age. In 40 per cent. of our series of 500 cases, the onset was before the age of thirty years. In the group with peri-articular changes predominating the age of onset is usually lower than in the group with special cartilaginous and bony changes.

**Sex.**—Among Garrod's cases there were 411 in women. Practically half of our series were males. The incidence as to sex is influenced by the inclusion of the cases of spondylitis, of which a large majority is in males. In women a close association with the menopause has been noted.

In America the incidence in the negro is relatively much less than in the white. Occupation and the station in life do not seem to have any special influence. Exposure to cold, wet and damp, errors in diet, worry and care, and local injuries are spoken of as exciting causes, but play only a predisposing part.

**INFECTION.**—The view is steadily gaining ground that the disease is due to infection and the evidence suggests certain varieties of streptococci as responsible. This seems more probable than that the disease is due to a specific organism. The *Streptococcus viridans* is the organism concerned in many cases. The arthritis is secondary to a focus of infection somewhere. The possible sources are many but infection of the mouth and throat probably takes first place. Abscesses about the teeth should always be searched for (X-ray study) and the tonsils carefully examined. Other sources are: infection of the nose or sinuses, pyorrhœa alveolaris, otitis media, chronic bronchitis, infection of the biliary or urinary tract, pelvic disease in women, and

infection of the prostate and seminal vesicles in men. The possibility of streptococcal infection from the intestinal tract must be considered although this is difficult to prove.

The acute onset, with fever in many cases, the polyarthritis, the presence of enlarged glands, the frequent enlargement of the spleen, the occurrence of pleurisy, endocarditis, and pericarditis in some cases, are all suggestive of an infection. The likeness of the lesions to those in arthritis from a specific cause, such as the gonococcus, is suggestive and also the association of the arthritis with definite foci of infection in many cases.

**METABOLIC.**—While the nutrition suffers in many cases there does not seem any evidence to support the view that the disease is primarily due to disturbance of metabolism. Pemberton has emphasized the lowered carbohydrate tolerance in some cases.

**Morbid Anatomy.**—The usual descriptions are of the late stages when extensive damage has occurred, for opportunities to study the early changes are few, although more frequent operations have extended our knowledge and skiagraphs have aided much. There are three main forms of change: (1) Lesions principally in the synovial membranes and peri-articular tissues (rheumatoid arthritis), (2) with atrophic changes in the cartilage and bones predominating, and (3) with hypertrophy and overgrowth of bone (osteo-arthritis). The first and second are most frequent in the joints of the extremities, the third in the spine. In many cases all forms of change are found, which speaks against the view that there are two distinct diseases. The changes in general are: (1) Effusion, which is not constant and shows no special features. (2) Changes in the synovial membrane. These are inflammatory and often hemorrhagic at the onset. There may be marked thickening and proliferation of the synovial fringes with the formation of villi—*villous arthritis*. (3) The capsule and surrounding tissues may be infiltrated and swollen. The peri-articular tissues show infiltration and swelling, and the enlargement of the joint is often due to swelling about it. (4) *Cartilage*. This may show erosion, ulceration, atrophy, or proliferation. The cartilage may disappear entirely, but the changes are often irregular and uneven and the cartilage may be replaced by fibrous tissue or by bone, the latter being most common at the edge of the cartilage. The cartilages may be soft and gradually absorbed or thinned (this often begins opposite the point of greatest involvement of the synovial membrane). (5) *Bone*. This may show atrophy of varying grade. If the cartilage is completely absorbed the surface of the bone often becomes hard and eburnated. In the *hypertrophic* form there is new bone formation which is most common at the edge of the articular surfaces. In the hip joint this may form an irregular ring of bone about the joint cavity. The commonest example of overgrowth of bone is seen in the so-called "Heberden's nodes," bony outgrowths at the terminal interphalangeal joints. There may be deposit of new bone in the ligaments, particularly in the spine, which may be converted into a rigid bony column. Proliferation of bone usually occurs at the margins of the joints in the form of irregular nodules—osteophytes. Bony ankylosis rarely occurs in the peripheral joints, but is common in the spine.

There may be extensive *secondary* changes. Muscular atrophy is common and may appear with great rapidity. Subluxation may occur, especially in the knee and finger joints. The hands often show great deformity, particu-

larly ulnar deflection. Contractures may follow and the joints become fixed in a flexed position. Neuritis and trophic disturbances may be associated; the neuritis is sometimes due to direct extension of the inflammatory process. Subcutaneous nodules occasionally occur.

The skiagraphs show the changes very well. Erosion of the cartilage is easily seen. In the type with predominant peri-articular changes the cartilage and bone often show little alteration. The occurrence of various changes in different joints or even in the same joint is common and bony change may occur with marked involvement of the peri-articular tissues.

**Symptoms.**—The onset may be acute or gradual. In the acute form a number of joints may be involved with high fever and the picture suggest rheumatic fever. In other cases the onset is acute in one joint and others are involved a few days later. With the gradual onset one joint is attacked and others follow. Some cases are between and may be termed subacute. In cases with an acute onset the attack may not persist very long; with the chronic onset the duration is usually prolonged. The acute onset occurs more frequently in the form in which peri-articular changes predominate.

**ARTHRITIS.**—In the acute form the joints are swollen, tender, and hot to the touch, but do not often show marked redness. There may be effusion in the larger joints. Pain is a marked feature and is increased by movement, the patient usually taking the position in which he has the greatest ease. When a joint is once attacked, the process does not subside quickly, and when the arthritis lessens some change remains in the joint which, however, may be very slight. The joints of the spine, especially in the cervical region, are often involved in the more acute forms, and in these there is rarely any permanent change. The temporo-maxillary joint is often involved, and arthritis here is always suggestive of this disease. The hands, when involved, show very characteristic changes. The knuckle joints are red, swollen, tender, and show limitation of motion. The fingers are often involved; swelling of the interphalangeal joints is common with a resulting thickening which gives a fusiform appearance. Partial dislocation, particularly at the terminal joint, is common. The knee-joints are often affected, with pain, effusion, limitation of motion, and later villous arthritis or subluxation. Thickening of the capsule usually occurs early.

In the *hypertrophic* (osteo-arthritis form) the process is rarely as acute as when the peri-articular parts are particularly involved (rheumatoid arthritis), but is usually polyarticular. The terminal finger joints, the hip joint, and the spine are especially affected. Pain is usually severe and the local features are not so marked. This form is more likely to be chronic.

**HEBERDEN'S NODES.**—These are small bony outgrowths ("little hard knobs"—Heberden) at the terminal phalangeal joints. They are much more common in women than in men. Heberden says "they have no connection with gout, being found in persons who have never had it," yet they are often regarded as indicating gout. In the early stage the joints may be swollen, tender, and slightly red, particularly when injured. The attacks of pain and swelling may come on at long intervals or follow injury. Sometimes they are the first manifestation of a general arthritis. Their distribution is not always regular and they are often largest on the fingers most used. They may be found in patients in whose larger joints the arthritis is of the other form.

The condition is not curable; but there is this hopeful feature—the subjects whose arthritis begins in this way rarely have severe involvement of the larger joints.

The MON-ARTICULAR FORM affects chiefly old persons, and is seen particularly in the hip and shoulder. It is identical with the general disease in its anatomical features. The muscles show wasting early and in the hip the condition ultimately becomes that described as *morbus coxæ senilis*. These cases not infrequently follow an injury. They differ from the polyarticular form in occurring chiefly in men and at a later period of life.

THE VERTEBRAL FORM (*Spondylitis*).—This may occur alone or with involvement of the peripheral joints. With an acute polyarthritis of the peripheral joints the spine may be involved, but there is usually no permanent change. With the *hypertrophic* form there is often bony proliferation and some spinal rigidity results which may involve the whole spine or only a part; in the latter case the lower dorsal and lumbar regions suffer most frequently. The condition may not involve more than a few vertebræ. The features are as variable as in the peripheral joints and there may be repeated acute attacks or a steady progressive process. In the general involvement the ribs may be fixed, the thorax immobile, and the breathing abdominal. There are two varieties of general involvement which are sometimes regarded as special diseases. In one (von Bechterew) the spine alone is involved, and there are pronounced nerve-root symptoms—pain, anæsthesia, atrophy of the muscles, and ascending degeneration of the cord. Von Bechterew thinks it begins as a meningitis, leads to compression of the nerve roots, loss of function of the spinal muscles, atrophy of the intervertebral disks, and gradually ankylosis of the spine. In the other—Strümpell-Marie type—the hip and shoulder joints may be involved (*spondylose rhizomélisque*), and the nervous symptoms are less prominent. Both are forms of arthritis deformans, and should neither be regarded nor described as separate diseases. *Spondylitis deformans* is more frequent in males, and trauma plays a part in its etiology. Local involvement is particularly common in the lumbar region and may cause sciatica and a great variety of referred pains. Pressure on the nerve-roots causes pain, paræsthesia, and muscle atrophy. Movement of the spine is usually restricted.

ARTHRITIS DEFORMANS IN CHILDREN.—Some cases resemble closely the disease in adults, in others there are very striking differences. In a variety described by Still, the general enlargement of the joints is associated with enlargement of the lymph glands and the spleen. The onset is usually before the second dentition, and girls are more frequently affected than boys. At first there is slight stiffness in one or two joints; others gradually become involved. The onset may be acute with fever or even with chills. The enlargement of the joints is due rather to peri-articular thickening than to bony change. The limitation of movement may be extreme, and there may be much muscular wasting. The enlargement of the lymph glands is striking, increases with fever, and may be general; even the epitrochlear glands may be as large as hazel nuts. The spleen can usually be felt. Sweating is often profuse and there may be anæmia, but heart complications are rare. The children look puny and generally show arrest of development.

GENERAL FEATURES.—*Temperature*.—In the acute attacks this may rise to 102° or 103° F., but is frequently lower and often persists for weeks with

a maximum about 100° F. The *pulse* is rapid in proportion to the fever, the most frequent range being from 90 to 110. Cardiac changes are found in a small proportion of cases. *Glandular enlargement* is common and may be general or especially marked in the glands related to the affected joints. The *spleen* is enlarged in some cases, the frequency being greater in the younger patients. *Subcutaneous nodules* occur in a few cases and are sometimes tender. The *blood* often shows a slight anæmia, not as marked as might be expected from the appearance of the patients. There is rarely much increase in the leucocytes and the differential count shows no peculiarity. The *urine* does not show any change of moment. The *skin* sometimes shows irregular areas of yellow pigmentation, especially on the face and arms. It may have a glossy appearance over the affected joints. Profuse sweating of the hands and feet is common. The *reflexes* are usually increased in acute cases and a return to normal is of good significance. They are sometimes absent. Muscular *atrophy* is common and sometimes advances very rapidly. It is most marked in the hands. Twitching of the muscles is not uncommon.

In some patients the bony atrophy is very marked. This is most common in females. In these disorganization of the joints occurs and the cartilage rapidly disappears. These cases usually progress rapidly downward. This atrophy is to be distinguished from that due to disuse.

**COURSE.—General Progressive Form.**—This occurs in two varieties, acute and chronic. The *acute* form may resemble, at its outset, rheumatic fever. There is involvement of many joints; swelling, particularly of the synovial sheaths and bursæ, but not often redness; there is moderate fever which is often persistent and may be from 99° to 100° F. for weeks. This pulse rate is usually high in proportion to the fever. In this form there may be repeated acute attacks, perhaps at intervals of years, or there may be repeated attacks in various joints. These usually leave definite changes, which may be slight at first, but tend to increase with subsequent attacks. Acute cases may occur at the menopause. Some cases progress very rapidly; they lose weight and strength; atrophy and arthritic deformity are marked; and they suggest a progressive septic process without suppuration.

The *chronic* form is more common, although many have an acute attack at some time, especially at the onset. The first symptoms are pain on movement and slight swelling, which may be in the joint itself or peri-articular. In some cases the effusion is marked, in others slight. The local conditions vary greatly, and periods of improvement alternate with remissions. At first only one or two joints are affected; gradually others are involved, and in extreme cases every joint in the body is affected. Pain is a variable symptom. Some cases proceed to the most extreme deformity without severe pain; in others the suffering is very great, particularly at night and during exacerbations of the disease. There are cases in which pain of an agonizing character is almost constant, quite apart from acute disturbances. Pain has an important influence in the production of deformity, as it hinders movement and the joints are kept in the position of greatest ease.

Gradually the shape of the joints is altered, partly by the thickening of the capsule and surrounding tissues, perhaps by osteophytes, and often by muscular contraction. Crepitus may be felt in the affected joint. Ultimately the joints may become immobile, not by a true bony ankylosis, although if

may be by the osteophytes which form around the articular surfaces, but more often from adhesions and peri-articular thickening. There is often an acute atrophy of the muscles and atrophy from disuse supervenes, so that contractures tend to flex the thigh upon the abdomen and the leg upon the thigh. Numbness, tingling, pigmentation or glossiness of the skin, and onychia may be present. In extreme cases the patient is helpless, and lies with the legs drawn up and the arms fixed. Fortunately, it often happens in these severe cases that the joints of the hand are not so much affected, and the patient may be able to knit or write, though unable to walk. In many cases, after involving two or three joints, the disease becomes arrested. A majority of the patients finally reach a quiescent stage, in which they are free from pain and enjoy fair health, suffering only from the inconvenience and crippling. Coincident affections are not uncommon. A small percentage show cardiac lesions, and the pulse rate is usually higher than normal.

**Diagnosis.**—The cases with an acute onset may be difficult to distinguish from *rheumatic fever*. The affected joints are rarely as tender as in rheumatic fever, and the smaller joints are more often involved. The presence of thickening in a joint, rapid muscular atrophy, a relatively high pulse rate in relation to the fever (in the absence of endocarditis), and the absence of response to salicylate medication speak against rheumatic fever. The diagnosis from *gonorrhoeal arthritis* may be difficult, but in this the small joints are usually not attacked so often, and after an onset with polyarthritis the majority of the affected joints clear, leaving one joint particularly involved. This rarely occurs in arthritis deformans. A careful search for gonococci is a great aid. In the chronic stage there may be difficulty in distinguishing this disease from *gout*. This is particularly marked in either disease without marked joint changes. The study of the skiagraphs is helpful and marked peri-articular changes speak for arthritis deformans. The finding of tophi or the estimation of the uric acid content of the blood may give the diagnosis of gout. It is important to distinguish *sub-deltoid bursitis* from the mono-articular form in the shoulder; the X-ray study is a great aid, as also in the recognition of disease of the *sacro-iliac* joint, *tuberculosis* of the *hip-joint* and spondylitis. Special importance attaches to the diagnosis of the spinal forms. There is no difficulty in the case of general involvement, but with local changes in the lower spine it is not so easy. Pain on and restriction of movement are important; the patient is careful to limit any motion of the spine. The root pains may be mistaken for evidence of visceral disease. Tuberculosis of the spine rarely offers any difficulty, especially with skiagrams.

**Prognosis.**—The age, general circumstances, character of the patient, the extent of arthritis, and the variety are all important. The outlook is not as dark as is usually described. If the source of infection can be found early and properly treated the prognosis is encouraging. In many patients the disease runs a certain course, and, if they can be brought through it with a minimum of damage, the ultimate outlook is good. In the form with peri-articular changes predominating, early diagnosis, treatment of the point of infection, the preservation of good nutrition, and a patient who is willing to fight are all encouraging factors. The outlook in the cases with the acute attacks is usually better than in those with a more chronic progressive course. Rapid muscular atrophy is of grave import. Cases in women beginning about the

menopause should always have a grave prognosis. Rapid advancement in the joint changes is serious. In the form in children the outlook is not good, but some recover entirely. The group with marked hypertrophic changes (osteo-arthritis) usually do well. Herberden's nodes are permanent, but in the larger joints it is rare for the condition to advance to absolute crippling, although there may be considerable interference with function. Spondylitis rarely advances to complete immobility of the whole spine. The outlook is good in the local cases, but depends somewhat on the occupation and possibility of trauma. The general condition is of importance and in those with marked nervous features the prognosis is not good.

**Treatment.**—Much depends on proper management and the pessimistic attitude is not justified. Too much stress can not be placed on the need of early diagnosis; the disease is often regarded as "rheumatic" and the treatment directed to this (especially restriction of diet and the giving of salicylates for long periods) is usually harmful.

**SOURCE OF INFECTION.**—Every effort should be made to find any such and prompt treatment carried out. Infection of the teeth and tonsils has always to be excluded. The possibility of infection of the bile passages should be considered. Whenever possible an autogenous vaccine should be prepared and used if the removal of the focus is not enough. Serums have not been of benefit in our experience.

**GENERAL MEASURES.**—The patient should be kept out of doors as much as possible and every effort made to improve the general health. The diet should be the most nourishing possible. The mistake of cutting down the proteins is often made. Regard must be had to the digestion, and it is more often the carbohydrates which should be reduced. Water should be freely given, as elimination is important. The bowels should be kept open, and for this the salines are useful. It is important to see that the patients are warmly clad in cold weather and guarded against chilling. *Hydrotherapy* is useful locally in the form of compresses, but the hot bath treatment, so often given, more frequently does harm than good, particularly in acute cases. Baths, when taken, should be of short duration. In more chronic cases bathing is sometimes of value. *Massage* is especially useful in the cases with synovial and peri-articular changes, and in them passive motion should be used early. *Climate* is of value in so far as patient is able to be out of doors and is saved from rapid changes of temperature.

**MEDICINAL.**—There is no drug which essentially influences the disease. The salicylates may aid in relieving pain, but should not be given for long periods. Iron, arsenic, and iodine are often useful. Iodine may be given as the tincture in doses of five to ten drops. Potassium iodide is sometimes of value when given for a long period. Thyroid and thymus gland extracts given persistently are sometimes beneficial. For the pain it is necessary to give drugs, although local measures should be used as much as possible. There are many which are available. Acetyl-salicylic acid (gr. x, 0.6 gm.), guaiacol carbonate (gr. v, 0.3 gm.), antipyrine (gr. iii, 0.2 gm.), and sometimes codeine (gr. ½, 0.03 gm.), are useful. Morphia should not be given on account of the danger of a habit.

**LOCAL.**—(a) Use of the joints must be governed by the condition. When the cartilage and bones are not involved, passive motion and massage are

useful, followed later by active motion. The patient should be taught simple exercises. When the cartilages and bones are involved, *rest* is usually advisable for a time. Every effort should be made to avoid contracture and displacement, and in this the use of splints during the night is often valuable. Caution should be exercised in advising complete fixation. This is sometimes useful for short periods but may result in fixation and is usually not advisable. (b) *Counter-irritation*. This is usually an aid, and the cautery, blisters, mustard, and iodine may be used. It is usually better to use light counter-irritation frequently than severe at longer intervals. (c) *Hyperæmia*. This may be active, and baking is a favorite method, but it should not be given for more than thirty minutes at a time. The temperature should be as high as the patient can stand. (d) *Hydrotherapy*. The persistent use of compresses is often of value. They may be put on in the evening and left on all night.

**SURGICAL MEASURES.**—These are useful for the correction of deformities. In villous arthritis operation is usually indicated. In the group with marked hypertrophy of bone removal of the outgrowths may be helpful.

**SPECIAL FORMS.**—(a) *Heberden's nodes*. Avoidence of irritation and injury is important, and in the case of pain the use of compresses is helpful. (b) *Spondylitis*. During the acute stages rest is essential and should be secured by a plaster jacket or simple apparatus. In the milder forms firm strapping may give relief. Trauma should be avoided. (c) *Knee-joint*. An elastic support is useful and may save the joint from injury.

**FOREIGN PROTEIN.**—This has proved useful in some cases, given intravenously as proteose (1-2 c.c. of a 4 per cent. solution) or as typhoid vaccine (75-150 millions). A sharp reaction is necessary if any benefit is to result.

**Arthritis Secondary to Acute Infection.**—While the majority of cases of arthritis are secondary to some infection, it is important to recognize various forms. (1) Those with a definite bacterial cause, such as gonorrhœal or tuberculous arthritis. These usually have fairly well defined features. (2) Those secondary to infections of doubtful etiology, such as typhus fever or measles. In some of these the arthritis is due to a secondary infection, but in others it appears to be due to the specific cause of the disease. (3) Arthritis secondary to infections in which there is no evidence of any organism in the joint. These are comparatively common and are difficult to designate. For example, arthritis, which may not be severe and subsides rapidly, occurs with an attack of tonsillitis. These have been termed "toxic" or "toxæmic" arthritis. The term "infectious" arthritis, sometimes applied, is not a satisfactory one. The cases in this group usually clear without leaving permanent damage, but if long continued they may result in the changes included under the heading of arthritis deformans.

**"Chronic Rheumatism."**—This term deserves mention because it is so commonly used, but its retention is not justified. There is no uniformity in its usage and it is applied without discrimination to all kinds of arthritis and frequently to conditions which have nothing to do with the joints. Painful conditions of the joints, muscles, fasciæ, bones, and nerves are all termed "rheumatism." There is no disease entity to which the term can be applied, and it would be an advantage to give it up entirely.

## II. INTERMITTENT HYDRARTHROSIS

The condition was described by Perrin in 1845 and only about 80 cases have been reported. It is characterized by a remarkable periodic swelling of one or several joints without fever. The knee is usually involved, sometimes the hip and wrist joints. The intervals between attacks vary from 2 to 30 days with an average of 12 days. The swelling may take place with great rapidity, and there may even be a sensation of water rushing into the joint. There are usually pain and stiffness. The periods may be from ten to twelve days, a month or even three months. Many of the cases have been in women and sometimes with marked hysterical symptoms. While some cases are secondary and represent a phase in the evolution of various articular lesions, there is a primary form characterized by a periodic swelling and nothing else. It is sometimes the joint equivalent to Quincke's oedema and may be associated with erythema, with angio-neurotic oedema, and in one of Garrod's patients with circumscribed oedema of the lips and eyelids. Some cases are due to anaphylaxis. A mother and daughter have been affected. The prognosis is not good; the attacks are apt to recur in spite of all treatment. Miller and Lewis obtained good results from the use of foreign protein (dead typhoid bacilli).

## C. DISEASES OF THE BONES

### I. HYPERTROPHIC PULMONARY OSTEO-ARTHROPATHY

**Definition.**—A symmetrical enlargement of the bones of the hands and feet, and of the distal ends of the long bones, occurring in association with certain chronic diseases, particularly affections of the lungs.

Bamberger in 1889 reported a condition of abnormal thickening of the long bones in bronchiectasis, and the next year Marie described other cases and named the condition.

**Etiology.**—*Clubbing* of the fingers, or the Hippocratic fingers, represent a minor manifestation of this condition. Many varieties occur; indeed, there is a monograph with sketches of some thirty or forty forms. It is met with perhaps most constantly in congenital disease of the heart, in tuberculosis and in other affections of the lungs, particularly bronchiectasis, in chronic jaundice, and in other chronic affections. In thoracic aneurism it may involve only the fingers of one hand. It usually comes on very slowly, but cases have been described of an acute appearance within a week or a fortnight. It may disappear. There is no bony alteration, but a fibrous thickening of the connective tissues with turgescence of the vessels. The condition is not easy to explain. The mechanical effect of congestion, the usual feature, explains the heart and lung cases, but not those of congenital syphilis and diseases of the liver. Others attribute it to a toxin.

Marie's syndrome is met with: (1) In diseases of the lungs and pleura, as in 43 of 55 cases collected by Thayer, and in 68 of Wynn's 100 cases.

Bronchiectasis is the most common, then pulmonary tuberculosis and empyema.

(2) Other affections, such as chronic diarrhoea, chronic jaundice, nephritis, and congenital syphilis. Marie regards the process as resulting from the absorption of toxins causing a periostitis; others have regarded it as a low form of tuberculous infection. The bones most frequently involved are the lower ends of the radius and ulna and the metacarpals, more rarely the lower end of the humerus, and the lower ends of the tibia and fibula.

**Symptoms.**—The affection comes on gradually, unnoticed by the patient. In other cases there is great sensitiveness of the ends of the long bones and of the fingers and toes. The fully developed condition is easily recognized. The hands are large, the terminal phalanges swollen, the nails large and much curved. Similar changes occur in the toes, and the feet look large, especially the toes and the malleoli. The bones of the fore-arms are diffusely thickened, particularly near the wrist, and the tibiae and the fibulae are greatly enlarged. Sometimes in advanced cases both ankles and knee-joints stand out prominently. The hypertrophy rarely affects the other long bones, though occasionally the extremities of the humerus and femur may be involved. The bones of the head are not attacked. Kyphosis may occur.

**Diagnosis.**—There is rarely any difficulty, as the picture presented by the hands and feet differs from that in acromegaly, and in practically all cases it is a secondary condition. *Treatment* should be directed to the primary disease; otherwise it is symptomatic.

## II. OSTEITIS DEFORMANS

(*Paget's Disease*)

**Definition.**—A chronic affection of the bones characterized by enlargement of the head, dorso-cervical kyphosis, enlargement of the clavicles, spreading of the base of the thorax and an outward and forward bowing of the legs.

The affection was described first by Sir James Paget, in 1877.

**Etiology.**—The generalized form is a rare disease, only 2 cases occurring among 20,000 medical cases at the Johns Hopkins Hospital. The etiology is unknown. Mother and daughter have been affected. Some have regarded it as luetic, others as due to arterio-sclerosis, which is a constant lesion. It may possibly be due to perversion of some internal secretion.

**Pathology.**—The skull, spine, and long bones are chiefly affected; those of the face, hands and feet are less involved. The skull may be as much as three quarters of an inch in thickness, and its circumference is increased. In one of Paget's cases it measured 71 cm. The shafts of the long bones are greatly thickened and they may weigh twice as much as a healthy bone of the same length. The femur is bent, the convexity forward; the tibiae may be huge and very much bowed anteriorly. The bones of the upper extremities are less often involved, the spine shows a marked kyphosis, sometimes partial ankylosis; the pelvis is broadened.

The process is a rarefying osteitis which gradually involves the centre of the bones. There is also new bone formation, both subperiosteal and myelogenous; the latter process gradually gains, and so the bones thicken.

**Symptoms.**—The disease begins, as a rule, in the sixth decade, sometimes with indefinite pains, but more frequently the patient notices first that the head begins to enlarge, so that he has to buy a larger hat. Then his friends notice that he is growing shorter, and that the legs are getting more and more bowed. There is a painful variety with great soreness of the arms and legs, which may be much worse at night. Headache, bronchitis and pigmentation of the skin, have been noted. The reduction in stature is very remarkable; one patient lost 13 inches in height.

**Diagnosis.**—The disease is readily recognized. The face differs from acromegaly, in which it is ovoid or egg-shaped with the large end down, while in Paget's disease the face is triangular with the base upward. In a few cases the disease may be limited to one or a few bones. There is a variety involving the tibiæ and fibulæ alone, and in some the femurs to a slight extent. These bones gradually enlarge, are bowed anteriorly and laterally, so that the only obvious features are a reduction in height with bowing of the legs. There is also a variety, sometimes known as tumor-forming osteitis deformans, in which the bones are much deformed with multiple hyperostosis and new growths. The relation of this to Paget's disease is doubtful.

**Treatment.**—With evidence of syphilis active treatment should be given. Otherwise symptomatic measures are indicated.

### III. LEONTIASIS OSSEA

In this disease there is hyperostosis of the bones of the cranium, and sometimes those of the face. The description is largely based upon the skulls in museums, but Allen Starr reported an instance in a woman, who presented a slowly progressing increase in the size of the head, face, and neck, the hard and soft tissues both being affected. He applied the term *megalcephaly* to the condition. Putnam states that the disease begins in early life, often as a result of injury. There may be osteophytic growths from the outer or inner tables, which in the latter situation may give the symptoms of tumor.

### IV. OSTEOGENESIS IMPERFECTA

(*Fragilitas Ossium, Osteopsathyrosis, Lobstein's Disease*)

**Definition.**—A prenatal and postnatal defective activity of the osteoblasts, rendering the bones abnormally brittle. It is often hereditary and is sometimes associated with a peculiar shape of the head and blue sclerotics.

**History.**—Lobstein described the disease as osteopsathyrosis in 1833, while Vrolik in 1849 described the prenatal idiopathic type as *osteogenesis imperfecta*. Recklinghausen and others thought the conditions were not identical, but the general opinion now seems to be that they are the same, and that *idiopathic fragilitas ossium*, whether prenatal or postnatal, is due to a deficient activity of the osteoblasts, whether in sub-periosteal or chondral ossification.

(Cameron). The terms *osteogenesis imperfecta congenita* and *tarda* best describe the two conditions.

**Etiology.**—Nothing is known except the single factor of heredity which occurs in a variable number of cases. Davenport and Conrad state that the heredity is typically direct and that the factor determining the irregular bone formation is a dominant one. The younger half of the family of an osteopathrotic parent will be affected, "but if neither parent, though of affected stock, has shown the tendency, then the expectation is that none of the children will have brittle bones." Biotypes occur, in some families the femur only, in some the humerus only is affected; and in some any pressure causes a break, while others show a much greater resistance.

**Symptoms.**—In the prenatal cases the child is often premature and still-born, the extremities short and thick with many fractures in all stages of healing. The head may feel like a crepitant bag of bones. The characteristic bitemporal enlargement has been described by Cameron. In the post-natal cases the onset is usually after infancy. While the liability to fracture, as a rule, decreases with years, it may persist until the age of fifty. Slight blows may cause a fracture, of which a child may have a score or more before puberty. As a rule, they are painless and heal readily.

**Treatment.**—Care should be taken to avoid fractures and, if they occur, every effort made to prevent deformity.

**Blue Sclerotics.**—Eddowes first described this remarkable condition which is more common in the inherited form. It may occur in individuals of the family who have never had fractures. It is not due to any color in the sclera itself but to increased transparency, possibly depending upon the absence of lime salts in the connective tissue.

## V. OSTEOMALACIA

### (*Mollities Ossium*)

This disease is characterized by pain, muscular weakness, and softness of the bones, due to decalcification, resulting in fractures and deformity. The great majority of the cases are in women; repeated pregnancies may play a part. A relationship to rickets is doubtful. Infection may have an influence. Disturbance of internal secretion, perhaps of the ovary, may be responsible. The onset is usually between the age of twenty and thirty. The bones are soft and show both decalcification and new formation. The earliest symptom is pain, especially in the back and sacral regions, increased by movement. Weakness is marked and there may be stiffness with contractures. The gait may be uncertain, sometimes spastic. The bony deformity is usually first in the spine or pelvis. Later there is marked deformity with fractures, callus formation and muscular wasting. The course is usually over some years and death may result from exhaustion or a terminal infection. The early diagnosis is difficult and may only be made when deformities appear. The X-rays are of value. In treatment, phosphorus in oil should be given (gr. 1/20-1/12, 0.003-0.004 gm.). The removal of the ovaries has been useful in some cases. Proper treatment for the symptoms should be given.

## VI. ACHONDROPLASIA

(*Chondrodystrophia Fetalis*)

**Definition.**—A dystrophy of the epiphyseal cartilages due to connective tissue invasion from the periosteum, in consequence of which the epiphyses and diaphyses are prematurely united and there is failure of the normal growth of the long bones. In consequence the subjects become dwarfs with normal heads and trunks, but short, stumpy extremities.

**Description.**—Achondroplastic dwarfs are easily recognized. They are well nourished and strong, and of average intelligence. Their height varies from 3 to 4 feet; the head and trunk are of about normal size, but the extremities are very short, the fingers, when the arms are at the sides, reaching little below the crest of the ilium. The important point is that in the shortness of the limbs it is the proximal segments which are specially involved, the humerus and femur being even shorter than the ulna and tibia (rhizomelia). The limbs are considerably bent, but this is more an exaggeration of normal curves and abnormalities in the joints than pathological curves. The features of rickets are absent. The hand is short and has a trident shape, since the fingers, which are of almost equal length, often diverge somewhat. The root of the nose is depressed, the back flat, and the lumbar lordosis abnormally deep, owing to a tilting forward of the sacrum. The scapulæ are short, the fibulæ longer than the tibiæ, and the pelvis is contracted; hence, the number of these cases reported by obstetricians. Heredity plays some part.

**Pathology.**—Anatomically it is a dystrophy of the epiphyseal cartilages, the cells of which are irregularly scattered, and the ground substances invaded by connective tissues from the periosteum, which sends in bands of tissues across the end of the diaphysis. The development of the bones with a membranous matrix seems normal.

Virchow described the disease as fetal cretinism, others as fetal rickets. Of late naturally its origin has been associated with disturbance of the pituitary function, or of its hormonal relations. On the other hand, Jansen of Leyden (1912) brings forward evidence to show that it results from a disturbance of the direct and indirect amniotic pressure, and brings it into relationship with a number of other fetal malformations. He states that the anatomical evidence is against changes in the sella turcica. But it is an argument in favor of some associated disturbance of the pituitary gland that achondroplasias often show precocious sexual development.

## VII. HEREDITARY DEFORMING CHONDRODYSPLASIA

(*Multiple Cartilaginous Erosions*)

The disease is characterized by the occurrence of multiple, usually symmetrical, cartilaginous or osteo-cartilaginous growths, generally benign, which result from proliferation and ossification of bone-forming cartilage, and cause bony deformities. The disease is hereditary in many instances; in one fam-

There were 26 cases in four generations. The American cases have been collected by Ehrenfried, who found that males were more often affected (3 to 1). The changes begin in infancy or early childhood, increase with skeletal growth and cease about the age of 22 years. The height is often less than normal. There are "irregular juxta-epiphyseal hyperostoses" most marked at the hips, knees, ankles, shoulders and wrists. There is often knock-knee and pes valgus, and the ulna may be relatively shortened. There may be symptoms due to pressure of the exostoses on nerves or vessels. Removal of the exostoses is indicated if they cause troublesome symptoms.

### VIII. OXYCEPHALY

**Definition.**—A cranial deformity associated with exophthalmos and impairment of vision.

**Description.**—The condition, known as *tower* or *steepleshead*, is characterized by great height of the forehead, sloping to a pointed vertex, with feebly marked supra-orbital ridges, and the hairy scalp may be raised above the normal level, looking as if perched on the top of a comb. The intelligence is unimpaired. The condition is usually present at birth, though in some instances it develops from the second to the sixth year. As this curious growth of the head proceeds, headache may be present, exophthalmos develops, and the vision becomes impaired, due to progressive optic atrophy. Smell is often completely lost. The deformity appears to be due to premature synostosis of certain sutures, notably the sagittal and coronal. As a result of the premature union of these two sutures the growth of the vault of the skull is restricted in both its antero-posterior and transverse diameters, and to accommodate the increasing bulk of the brain a compensatory increase in height takes place. Eventually the anterior fontanelle closes, but there is reason to think that this occurs at a later date than the normal, and its former site is marked by a slight protuberance with thinning of the bone. (Morley Fletcher, *Quarterly Jour. Med.*, IV., 1911.)

The optic neuritis and atrophy are the result of pressure exerted by the growing brain and may be compared to that of cerebral tumor. As yet we do not know the cause of this premature synostosis. Rickets and syphilis are not responsible. The condition is one for which a decompression operation with ventricular puncture is indicated.

### IX. HEREDITARY CLEIDOCRANIAL DYSOSTOSIS

This is a familial and hereditary disease in which the skull is large and the fontanelles and sutures close late. There may be enlargement of the large fontanelle. There are prominent cranial bosses. The clavicles are defective, the acromial end usually being absent. There may be pseudo-arthritis or absence of a part or the whole of the clavicles. The mobility of the shoulders is increased and they can be made to meet under the chin. Many of the patients show genu valgum, hereditary and familial, transferred through either parent.

*Dr. Bon Behari Bose*  
Uttarpara, Hooghly.

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